

Evaluation of primary retroperitoneal masses by computed tomography scan

Anantkumar Chaudhari¹, Purvi D Desai², Mahesh K Vadel², Kumarbhargav Kaptan³

¹Consultant Radiologist, Concept Imaging, Surat, Gujarat, India.

²Department of Radiodiagnosis and Imaging, Government Medical College, Surat, Gujarat, India.

³Consultant Pathologist, Doctor's Lab, Surat, Gujarat, India.

Correspondence to: Anantkumar Chaudhari, E-mail: anantchaudhari85@gmail.com

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Abstract

Background: Retroperitoneum is one of the largest spaces in the body where the primary and metastatic tumors grow silently to large sizes before the clinical signs and symptoms appear. Though retroperitoneal masses are rare with reported incidence of 0.3% to 3.0%, majority of them are malignant. Early and accurate diagnosis is crucial. Of the many diagnostic modalities, radiological investigation such as computed tomography (CT) is the modality of choice for the primary evaluation.

Objective: To evaluate the usefulness of CT scan in evaluation of the retroperitoneal masses.

Materials and Methods: Total 30 cases of retroperitoneal mass were studied by CT scan using oral and intravenous contrast in the Department of Radiodiagnosis and Imaging, Government Medical College and New Civil Hospital, Surat, Gujarat, India, from July 2010 to July 2013.

Result: Of the 30 cases, 17 were male and 13 were female patients. A total of 63.3% of the lesions were malignant and 36.6% of the lesions were benign. Retroperitoneal lymphoid masses were the most common.

Conclusion: CT scan is safe, simple, minimally invasive, and a cost-effective method in the diagnosis of primary retroperitoneal masses. It is highly effective in determining the exact location, extension, metastatic lymphadenopathy, vascular invasion, and distant metastases with fair accuracy. It is a vital guide in the procedures such as guided FNAC and biopsy. It provides important clues for the final diagnosis. Thus, CT is the modality of choice in the evaluation of retroperitoneal masses. However, differentiating the malignant lesions and metastatic lymph nodes from each other is difficult on CT features alone.

KEY WORDS: Retroperitoneal masses, CT scan, primary, evaluation

Introduction

Retroperitoneal space is limited anteriorly by peritoneal covering (parietal peritoneum), posteriorly by posterior abdominal wall (transversalis fascia), superiorly by 12th rib and vertebrae, inferiorly by the base of sacrum and iliac crest,

and laterally by the sites of borders of quadratus lumborum muscles. It extends from diaphragm superiorly to the level of the pelvic viscera inferiorly. It contains connective tissue, kidney, adrenals, ureter, aorta with its branches, inferior vena cava with its tributaries, and lymph nodes.^[1] Retroperitoneum is divided into three spaces, at the level of kidneys by anterior (Gerota's fascia) and posterior renal fascia (Zuckerkind's fascia). These are (1) anterior pararenal space, (2) perirenal space, and (3) posterior pararenal space.^[2]

For many years, the retroperitoneum was a difficult region to image radiographically. The signs and symptoms of retroperitoneal disorders are frequently vague and poorly localized. With the advent of ultrasonography (USG), computed tomography (CT), and magnetic resonance imaging (MRI), retroperitoneal disorders are readily identified and their spreads through various retroperitoneal compartments are assessed.

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Primary retroperitoneal masses are rare, with reported incidence of 0.3% to 3.0%. Majority of them (70%–85%) are malignant and 15%–25% are benign.^[3] A total of 75% are mesenchymal in origin, 24% neural in origin, and 1% from embryonic rest. They usually present in middle aged, but can occur at any age group.

Primary retroperitoneal masses mainly comprise the following:

- Lymph node masses,
- Malignant primary retroperitoneal masses, and
- Benign primary retroperitoneal masses.

They grow aggressively and at the time of diagnosis, they invade the surrounding structures. Lymphadenopathy and distant metastases are rare in primary retroperitoneal malignancies. Radiological investigation such as CT is the modality of choice for the evaluation of primary retroperitoneal masses. Disease processes confined to the retroperitoneum, including the primary retroperitoneal tumors, retroperitoneal fibrosis, retroperitoneal collections, hemorrhages, abscesses and abnormalities related to lymph nodes, and lesions of the retroperitoneal organs.

The first step is to confirm whether the tumor is located within the retroperitoneal space. It is useful to observe the displacement of normal anatomic structures.^[4,5] Anterior displacement of retroperitoneal organs (e.g., kidneys, adrenal glands, ureters, ascending and descending colon, pancreas, and portions of the duodenum) strongly suggests that the tumor arises in the retroperitoneum. So the displacement of these vessels can be helpful as well. Before a tumor can be described as primarily retroperitoneal, the possibility that the tumor originates from a retroperitoneal organ must be excluded. Some radiologic signs that are helpful in determining tumor origin include the “beak sign,” the “phantom (invisible) organ sign,” the “embedded organ sign,” and the “prominent feeding artery sign.”^[5,6] When there is no definite sign that suggests an organ of origin, the diagnosis of primary retroperitoneal tumor becomes likely.

Beak Sign—when a mass deforms, the edge of an adjacent organ into a “beak” shape, it is likely that the mass arises from that organ (beak sign). On the other hand, an adjacent organ with dull edges suggests that the tumor compresses the organ but does not arise from it.^[5,6]

Phantom (Invisible) Organ Sign—when a large mass arises from a small organ, the organ sometimes becomes undetectable. This is known as the phantom organ sign. However, false-positive findings do exist, as in cases of huge retroperitoneal sarcomas that involve other small organs such as the adrenal gland.^[5,6]

Embedded Organ Sign—when a tumor compresses an adjacent plastic organ (e.g., gastrointestinal tract, inferior vena cava) that is not the organ of origin, the organ is deformed into a crescent shape. In contrast, when part of an organ appears to be embedded in the tumor it is negative embedded organ sign.

The tumor is in close contact with the organ and the contact surface is typically sclerotic with desmoplastic reaction. Occasionally, the contact surface becomes ulcerative. When the embedded organ sign is present, it is likely that the tumor originates from the involved organ.^[5,6]

Prominent Feeding Artery Sign—hyper vascular masses are often supplied by feeding arteries that are prominent enough to be visualized at CT or MRI, a finding that provides an important key to understanding the origin of the mass.^[5,6]

Familiarity with the specific features of various retroperitoneal tumors often allows accurate histologic diagnosis and helps suggest proper management. Like few important points to be remembered: (1) some retroperitoneal tumors have specific patterns of growth and extension that aid in narrowing the differential diagnosis, (2) some tumors grow and extend into spaces between preexisting structures and surround vessels without compressing their lumina (e.g., lymphangiomas and ganglioneuromas). Another entity with this growth pattern is lymphoma. This neoplasm tends to surround adjacent vessels, manifesting with the “CT angiogram sign” or “floating aorta sign.” (3) Few tumors such as tumors of the sympathetic ganglia (e.g., paragangliomas, ganglioneuromas) tend to extend along the sympathetic chain and have an elongated shape.^[5,7] (4) Some tumor contents can be clearly demonstrated at CT and MRI and provide strong clues that help narrow the differential diagnosis such as presence and absence of fat. A mass that is homogeneous and well-defined and consists almost entirely of fat represents lipoma.^[7] When the mass is somewhat irregular and ill-defined but contains fat, the diagnosis of liposarcoma should be considered. Liposarcomas are the most common sarcomas of the retroperitoneum.^[8,9] Teratomas are also characterized by the presence of fat, and mature teratomas can be characterized by the presence of fluid attenuation or signal intensity, fat–fluid levels, and calcifications.^[4] A limited number of tumors commonly contain myxoid stroma, which helps to narrow the differential diagnosis. Myxoid stroma appears hyperintense on T2-weighted MR images and shows delayed enhancement after injection of contrast medium.^[10] Tumors that commonly contain myxoid stroma include neurogenic tumors (schwannomas, neurofibromas, ganglioneuromas, ganglioneuroblastomas, malignant peripheral nerve sheath tumors), myxoid liposarcomas, and myxoid malignant fibrous histiocytoma.^[10] Tumors that less commonly contain myxoid stroma include desmoid tumors, hemangiopericytomas, leiomyomas, leiomyosarcomas, malignant pericytomas, rhabdomyosarcomas, and malignant mesenchymomas.^[10] (5) Necrotic portions within tumors have low attenuation without contrast enhancement at CT. Necrosis is usually seen in tumors of high-grade malignancy such as leiomyosarcomas.^[4] Extremely hypervascular tumors such as paragangliomas sometimes contain hemorrhagic necrosis and manifest with fluid–fluid levels. (6) Some tumors are completely cystic in appearance. These include lymphangiomas and mucinous cystic tumors. Solid tumors with a partially cystic portion

include neurogenic tumors.^[8] (7) Lymphomas are the most commonly encountered tumors composed of small round cells. They are homogeneous with minimal contrast enhancement at CT. (8) Extremely hypervascular tumors include paragangliomas and hemangiopericytomas. Moderately hypervascular tumors include myxoid malignant fibrous histiocytomas, leiomyosarcomas, and many other sarcomas. Hypovascular tumors include low-grade liposarcomas, lymphomas, and many other benign tumors.^[10]

Thus, it showed that CT scan is the vital primary mode of assessment in the evaluation of primary retroperitoneal masses, which is necessary even for further assistance in guided FNAC and biopsy analysis. Though it provides very useful clues for final diagnosis, even CT scan alone may not be sufficient for accurate diagnosis. Further confirmatory investigations such as FNAC and biopsy would be necessary for diagnosis.

Materials and Methods

This prospective study "Computerized Tomographic study of retroperitoneal masses" was carried out in the Department of Radiodiagnosis and Imaging, Government Medical College and New Civil Hospital, Surat, Gujarat, India from July 2010 to July 2013. Patients suspected to have retroperitoneal masses clinically were further evaluated with CT. As the procedure was provided free of cost, a total of 30 patients were evaluated by CT scan, which was granted throughout the period. Whenever possible, patients were further evaluated by fine needle aspiration cytology, biopsy, and/or other operative procedure for comparison. The CT examination was performed on Somatom Emo 6 Siemens machine. Patients were first scanned in supine position and relaxed expiration phase. Noncontract study was followed by the intravenous contrast study. Preprocedure preparations include patients should be nil by mouth for about 6 h before conducting study. Ionic or nonionic contrast agents were used for intravenous contrast studies. The contrast agents routinely used were 75% urografin or Iopamidol (iopamiro, lek pomidal) or Iohexol (Omnipaque). Approximately 80 mL of contrast was administered followed by conduction of contrast sensitivity test in each patient. Oral contrast (urografin) was also given to each patient for opacification of gastrointestinal tract. Scans were obtained in hepatic and portal phase and delayed scans were also obtained in some cases. Delayed scans were obtained whenever necessary.

Scanning protocol: region from both domes of diaphragm to pubic symphysis was included. Patient position was supine with arms above head. Scanogram was fixed for anteroposterior length of 512 mm. Scan parameters included spiral mode, slice thickness: 6 mm, feed: 12 mm, pitch: 1.5, scan orientation: craniocaudal, scan delay: 45 s, voltage: 120 kV and 200 mA, FOV: 350 mm, filter: AB50, window setting: 300/40 for soft tissue and 1500/300 for bone window.

Postprocessing: Multiplanar reconstruction and spiral reconstruction of images was done to obtain scans as thin as 2 to

3 mm in thickness, whenever necessary. All examinations were evaluated for the exact location, extent, local invasion vascular encasement, invasion, displacement of adjacent structures, and distant metastases. USG-guided fine needle aspiration biopsy or true cut biopsy was carried out in some patients. CT diagnosis of these lesions was made and confirmation was obtained by fine needle aspiration biopsy, open biopsy, or postoperatively. Patients were categorized according to the type of lesion. The data from these studies were tabulated and used to study the efficiency of the CT as a diagnostic modality.

Result

Total number of patients studied was 30. Of these, 17 (56.6%) were male and 13 (43.3%) were female patients. A total of 63.3% of the lesions were malignant and 36.6% of the lesions were benign. CT examinations were done using oral and intravenous contrast in all patients. Commonly affected age group was 6th, 5th, and 3rd decade followed by 7th and 4th decade. Youngest patient was aged 6 months whereas the oldest was of 65 years [Tables 1 and 2].

Retroperitoneal lymph node masses were the most common malignant lesions. Renal cell carcinoma and pancreatic adenocarcinoma were the next most common lesions. Benign lesions were equal in male and female patients. In this study, retroperitoneal lymphadenopathy was the most common

Table 1: Age and sex distribution

Age (years)	Males	Females	Total
0-1	—	2	2
2-10	1	2	3
11-20	1	1	2
21-30	1	4	5
31-40	4	—	4
41-50	4	1	5
51-60	2	3	5
61-70	4	—	5
Total	17	13	30

Table 2: Sex distribution in malignant lesions

Diseases (n = 30)	Males	Females	Total
	No. of cases	No. of cases	
Retroperitoneal lymph node mass	8	2	10
Renal cell carcinoma	4	1	5
Neuroblastomas	—	1	1
Paragangliomas	—	1	1
Pancreatic adenocarcinoma	1	2	3
Wilm's tumor	—	2	2
Total	13	9	22

lesion. The retroperitoneal lymph nodes were involved by the primary neoplasm, from lymphomas and metastatic tumors. Most of the lesions showed heterogeneous enhancement. Retroperitoneal lymphoma showed mild enhancement; whereas metastatic lymph node mass showed heterogeneous postcontrast enhancement. Fatty component was present in renal angiomyolipoma and adrenal adenoma. Calcifications were present in renal cell carcinoma, lymph node mass, and in hydatid cyst. Necrotic / hemorrhagic components were seen in metastatic lymph node mass followed by renal cell carcinoma and pancreatic adenocarcinoma. Vascular encasement was noted in a total of 11 cases (36.6%). Local invasion was seen in eight cases (26.6%). Metastatic testicular lymph nodes are more common [Tables 3 to 7].

In this study, six (20%) cases were of lymphoma of which two (33.3%) were of Hodgkin's disease, and four (66.6%) were NHL. All the cases in this study had stage IV disease. CT was highly accurate in staging the lymphomas. Extranodal sites of involvement were found in one case of NHL. One of the extra

nodal sites was liver. We have investigated nine (30%) cases of lymph node metastases from other malignancies, four (13.3%) were from testicular origin, (6.6%) from renal malignancies, one (3.3%) from pancreatic adenocarcinoma, Wilm's tumor, and paraganglioma. Most of the nodal masses showed mild enhancement in 1 case, heterogeneous enhancement in 18 cases, and moderate enhancement in 1 case. Retrocrural nodes were present in one case of non-Hodgkin's lymphoma. One male patient of 30 years age, an operated case of teratocarcinoma testes, post chemotherapy came for follow-up CT scan. CECT showed regression in the nodal mass size [Tables 8 and 9].

Discussion

In the late 1960s and early 1970s, the various compartments of the retroperitoneum were studied by using three compartment models.

Table 3: Sex distribution in benign lesions

Diseases	Sexes		Total
	Males	Females	
	No. of cases	No. of cases	
Renal abscess	2	—	2
Retroperitoneal lymphangioma	1	—	1
Renal hydatid cyst	—	1	1
Renal angiomyolipoma	—	2	2
Adrenal adenoma	—	1	1
Adrenal myelolipoma	1	—	1
Total	4	4	8

Table 4: Enhancement pattern on CT

Diseases (n = 30)	Nonenhancing hypodense	Enhancement			
		Mild	Moderate	Intense	Heterogeneous
Retroperitoneal lymph node mass	—	1	1	—	8
Renal cell carcinoma	—	—	1	—	4
Renal abscess	—	2	—	—	—
Neuroblastomas	—	—	—	—	1
Paragangliomas	—	1	—	—	—
Pancreatic adenocarcinoma	—	—	1	—	2
Renal angiomyolipoma	—	2	—	—	—
Renal hydatid cyst	—	1	—	—	—
Wilm's tumor	—	1	—	—	1
Adrenal adenoma	—	—	1	—	—
Adrenal myelolipoma	—	1	—	—	—
Lymphangioma	1	—	—	—	—
Total	1	11	4	—	16

CT, computed tomography.

Table 5: Fatty component and calcifications on CT

Diseases (n = 30)	Fatty component		Calcifications	
	No. of cases	Out of 30	No. of cases	Out of 30
Renal angiomyolipoma	2	6.6%	—	—
Adrenal adenoma	1	3.3%	—	—
Renal cell carcinoma	—	—	1	3.3%
Retroperitoneal lymph node mass	—	—	1	3.3%
Renal hydatid cyst	—	—	1	3.3%
Wilm's tumor	—	—	1	3.3%
			-	

CT, computed tomography.

Table 6: Necrotic/hemorrhagic components on CT

Diseases (n = 30)	Necrosis/hemorrhagic component	
	No. of cases	Out of 30
Retroperitoneal lymph node mass	5/-	16.6%
Renal cell carcinoma	4/-	13.3%
Renal abscess	-/1	3.3%
Neuroblastomas	-/-	-
Paragangliomas	-/-	-
Pancreatic adenocarcinoma	3/-	10%
Renal angiomyolipoma	-/-	-
Renal hydatid cyst	-/-	-
Wilm's tumor	2/-	6.6%
Adrenal adenoma	-/-	-
Adrenal myelolipoma	-/-	-
Lymphangioma	-/-	-

CT, computed tomography.

Table 7: Vascular encasement / invasion and local invasion

Diseases (n = 30)	Vascular encasement/invasion		Local invasion	
	No of case	Out of 30	No of case	Out of 30
Retroperitoneal lymph node mass	5	16.6%	2	6.6%
Renal cell carcinoma	3	10%	2	6.6%
Renal abscess	—	—	1	3.3%
Pancreatic adenocarcinoma	1	3.3%	1	3.3%
Wilm's tumor	1	3.3%	1	3.3%
Lymphangioma	1	3.3%	—	—

Table 8: Lymphadenopathy and distant metastases in primary lesions

Diseases	Lymphadenopathy		Distant metastases	
	No. of cases	Out of 30	No. of cases	Out of 30
Wilm's tumor	1	2	—	—
Pancreatic adenocarcinoma	1	3	1	3
Renal cell carcinoma	2	5	1	5
Neuroblastomas	—	—	1	1
Paraganglioma	1	1	—	—
Total	5	11	3	9

Stephens et al.^[11] studied 19 patients of which there were 12 male and 7 female patients. Mean age ranged from 2 to 72 years. A total of 18 cases had malignant lesions and a single case was of benign cystic lesion. They concluded that CT has a major role in the diagnosis of retroperitoneal tumors and their

Table 9: Retroperitoneal lymph node masses (metastatic)

Diseases	Males	Females	Total
Lymphoma	4	2	6
Ca testis	4	—	4
Renal cell carcinoma	1	1	2
Pancreatic adenocarcinoma	1	—	1
Wilm's tumor	—	1	1
Paraganglioma	—	1	1
Total	10	5	15

recurrences. Even in the cases of advanced tumors, the knowledge provided by CT is invaluable in developing a rational approach for its management. In this study, of the 30 patients, 17 (56.6%) were male and 13 (43.3%) were female patients. Of the 30 cases, 19 (63.3%) had malignant and 11 (36.6%) had benign lesions. Malignant lesions were slightly more common in male patients (12 [40%]), whereas benign ones were slightly more common in female patients (6 [20%] cases). These findings are similar to that of Stephens et al.^[11]

Jones et al.^[12] studied 40 patients with lymphoma with the CT of chest and abdomen. CT has particular utility for the assessment of lymphomatous involvement of mesenteric lymph nodes, high retroperitoneal lymph nodes, and extranodal abdominal sites other than spleen or liver (e.g., pancreas, kidneys). Abdominal CT is a useful noninvasive diagnostic technique for the initial staging and subsequent evaluation of selected patients with lymphoma. Zelch and Haaga^[13] studied the advantages of CT over lymphography. CT provides a better means for assessing the true extent of disease and permits accurate follow-up assessment of therapy. CT examination does not require significant technical expertise. CT scan is an excellent means for guiding biopsy procedures even for retroperitoneal abnormalities. The most significant disadvantage of CT is its inability to resolve or detect neoplastic disease within normal-sized lymph nodes. Alymlahi et al.^[14] bilateral adrenal neuroblastomas the bilateral neuroblastoma is a rare entity. Medical imaging is very helpful in the diagnosis and the staging of this tumor. Hirasaki et al.^[15] concluded that neuroendocrine tumor should be included in the differential diagnosis of a retroperitoneal mass, although composite paraganglioma-ganglioneuroma in the retroperitoneum is very rare.

Hayes et al.^[16] studied 28 patients with CT retrospectively and found that CT features were correlated with clinical and pathologic findings. There were 16 men and 12 women and the average age was 37 (range: 11–70 years). A total of 24 patients (86%) had hypertension. Four patients (14%) had malignant paragangliomas. They classified them by location as suprarenal (26%), renal hilar (32%) or infrarenal (42%). The suprarenal paragangliomas could not be distinguished from the ipsilateral adrenal gland on CT. They concluded that the average size of the functional tumor was smaller (7 cm) than that of the nonfunctional tumors (12 cm), smaller tumors were

more likely to be homogeneous and had well-defined margins. Their findings indicated that extra-adrenal retroperitoneal paragangliomas were functionally active more often than it was previously reported; however, no CT features are unique for paragangliomas. Storm and Mahvi^[17] stated that the diagnosis of primary retroperitoneal masses is approached best by CT scan-guided trans-retroperitoneal core biopsy or by open biopsy. They concluded that excisional or wedge biopsy is the preferred method for undiagnosed lymphadenopathy. Sklair-Levy et al.^[18] have conducted 267 biopsies by using cutting needles. CT-guided aspiration core biopsies were sufficient to establish a diagnosis in lymph proliferative disorders in 82.5% of the cases. In the light of this experience, we suggest that imaging-guided core needle biopsy be used as the first step in the workup of many patients with lymphoma. They obtained positive results diagnostic in 17 (73.9%) and nondiagnostic in 6 (26%). Hugosson C et al.^[19] reviewed 31 children with abdominal neuroblastomas (median age 2 years) with USG and CT. They observed that CT and MRI were superior to USG. There is no significant difference between CT and MRI, in the assessment of the location or size of tumor. Intraspinal extension was more distinctly demonstrated with MRI. They concluded that local disease was best assessed by either CT or MRI, whereas metastatic disease was best revealed by CT and that imaging may be valuable for clinical assessment and pretreatment staging of abdominal neuroblastomas.

Madrigal Rubiales et al.^[20] studied two cases of extra-adrenal nonsecretory retroperitoneal paragangliomas. Both were men. They studied both with USG, CT, and MRI. They showed solid homogeneous retroperitoneal masses, intimately adhered to aorta and located in left upper abdomen. They concluded that extra-adrenal retroperitoneal paragangliomas are rare tumors, particularly the nonsecretory. The only reliable criteria for malignancy were extensive capsular invasion and distant metastases.

Yang et al.^[21] have classified cystic masses of the retroperitoneum as either neoplastic or non-neoplastic. CT may provide important information regarding lesion location, size, and shape; the presence and thickness of a wall; the presence of septa, calcifications, or fat; and the involvement of adjacent structures.

Now the various studies abovementioned had their observations, which add up in the experience. This study also adds up to the continual progress in CT analysis of retroperitoneal masses.

Conclusion

As primary retroperitoneal masses are uncommon, many different studies should accumulate their experience before many different findings can be considered characteristic for one particular tumor and which will be of useful for future enhanced diagnosis. This study also points that it is necessary to first distinguish whether it is primary or other and enumerate the usefulness of CT in the evaluation of primary retroperitoneal masses, which is sometimes the vital

procedure for guided FNAC and biopsy to reach at the final diagnosis. This study also states that the primary retroperitoneal masses were less common than the retroperitoneal lymph node enlargements and metastatic lesions. However, it is not possible always to differentiate it by CT scan alone, it requires constellation of clinical information along with other investigations and specific features. CT plays an important role in the detection of lymphomatous involvement of nodes, extent of lesion, and in the detection of extranodal sites of involvement. CT also plays important role in the follow-up after radiotherapy or chemotherapy. Sensitivity of CT versus lymphography in demonstrating NHL is reported as 86% for CT and 100% for lymphography, with an overall accuracy of 82% and 91%, respectively. CT not only evaluates lymphadenopathy but most of the times it shows evidence of primary lesions while evaluating retroperitoneal lymphadenopathy. The basic purpose of this study is to share the data about the CT findings and its usefulness in the evaluation of retroperitoneal masses. With the advent of newer modes of investigations, the scenarios will surely change in the future and more and more such studies are needed for establishing the various criteria.

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