Lipomatous Hemangiopericytoma: A Rare variant of Hemangiopericytoma

Dr.Tarang Patel[1], Dr.Priti P Trivedi[2], Dr. Divya Kriplani[3], Dr.Manoj J. Shah[4]

ABSTRACT

Introduction: Lipomatous hemangiopericytoma is a very rare variant of hemangiopericytoma. Histologically it is characterized by the presence of hemangiopericytomatous vasculature and mature adipocytes. **Case report:** We describe a Lipomatous hemangiopericytoma that arose retroperitoneally in a 45 year old male. **Conclusion:** It is important to differentiate these benign neoplasm's from other tumors showing a dual adipocytic and spindle-cell composition that have a more aggressive outcome.

Key words: hemangiopericytoma variant, Immunohistochemistry, lipomatous, liposarcoma

[1] Resident, [2] Professor, [3] Fellow, [4] Professor & Head,
Department of pathology, The Gujarat cancer and research institute, Ahmedabad, Gujarat, India
Corresponding author mail: tarangpatel_86@yahoo.co.in

Conflict of interest: None

INTRODUCTION

In 1995 Nielson et al [1] reported a series of three previously undescribed soft tissue tumors, composed of an admixture of hemangiopericytoma and mature adipose tissue. They coined the term Lipomatous hemangiopericytoma for these neoplasms. Lipomatous hemangiopericytoma (LHPC) was formally described as a new entity in the 2002 edition of the WHO classification of soft tissue tumors, as a rare hemangiopericytoma variant composed of mature adipocytes and hemangiopericytoma like areas. [2] We report a case of this rare tumor in a 45 year old man.

CASE REPORT

A 45-year-old male presented with complaint of abdominal swelling for past two years in July 2010. Patient was hypertensive but had no other significant past or family history. An ultrasound (USG) abdomen and pelvis revealed the presence of large retroperitoneal solid lesion with few internal cystic areas. A CT scan thorax abdomen and pelvis showed 27x20x14 cms Isodense to hypodense lobulated mass lesion

intraperitoneally in the midline, extending to either side. Lesion extended from level of upper pole of right kidney up to the pelvic cavity inferiorly. Post contrast significant enhancement of lesion was noted. The liver, gall bladder, spleen and pancreas were normal. Bilateral kidneys were normal. There was no lymphadenopathy. The rectum, sigmoid colon, prostate and seminal vesicles were all normal. Thorax was normal. Patient underwent surgery for the resection of mass in October 2011.

On gross examination, the mass weighed 5 Kg and measured 36x30x10 cms. The outer surface was encapsulated. On cut section the mass had a lobulated surface with solid and cystic components. The solid part showed a myxoid to yellowish surface with soft to firm consistency.

Microscopic examination showed, spindle cells arranged haphazardly around variable sized, thin walled blood vessels, which also showed a typical staghorn pattern (Fig. 1C & 1D). Cells had small round to oval nuclei, fine granular chromatin and ill-defined cell borders (Fig. 1E). Closely admixed foci of mature adipose tissue were seen throughout the tumor (Fig.1A,1B & 1F). Stromal fibrosis was evident. Mitoses were not seen.

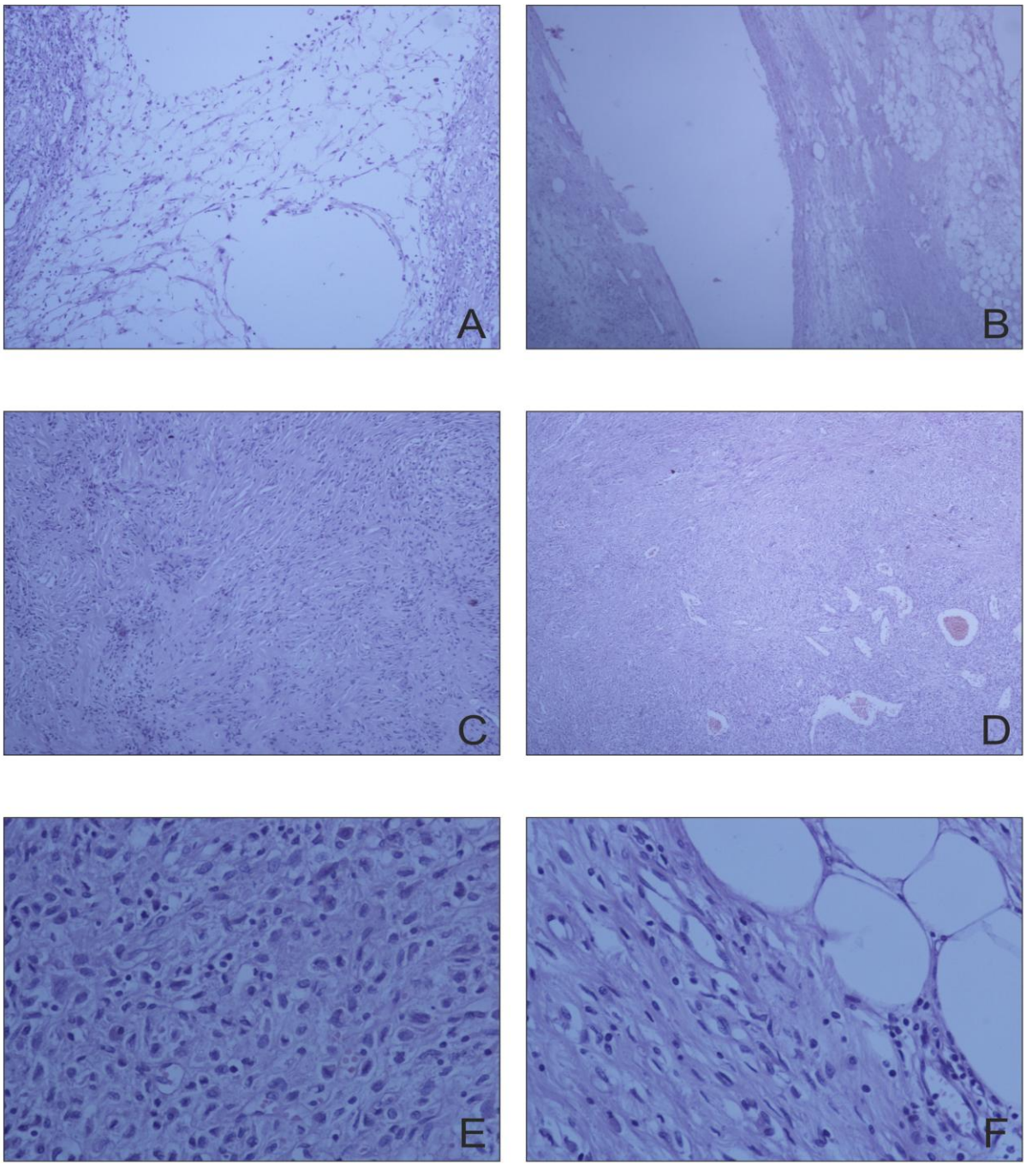
A
B
C
D
E
F

Figure 1: Lipomatous hemangiopericytoma. (A&B) photmicroghaph shows Mature adipose tissue (arrow) intermingled with spindle cells, 4X. (C&D)Cellular areas of HPC with focal areas showing branching vascular pattern (arrowhead) 4X. (E) High power view of the tumor showing oval to round cells which lack pleomorphism 40X. (F) A typical region of hemangiopericytoma adjacent to a fat rich component of the tumor 40X.

Immunohistochemistry was performed and showed strong and diffuse positivity for vimentin, CD 34, CD 99 and bcl-2 (Fig. 2A,2B & 2C). Cells were negative for S-100 (Fig. 2D), Actin and CD117.

Based on the morphological and immunohistochemical findings a final diagnosis of lipomatous variant of hemangiopericytoma was made.

A follow up CT scan done one year later revealed a 6.4x5.8 cm sized lesion in the pelvic cavity on the left side. Radiological possibility of postoperative collection / residual tumor was given. Biopsy showed only blood clot. Patient was kept on regular follow-ups. Subsequent scans done at six month and one-year interval showed a spontaneous decrease in the size of the lesion to 2.5 cm and 1.8 cm respectively. The patient is well and on regular follow up till date.

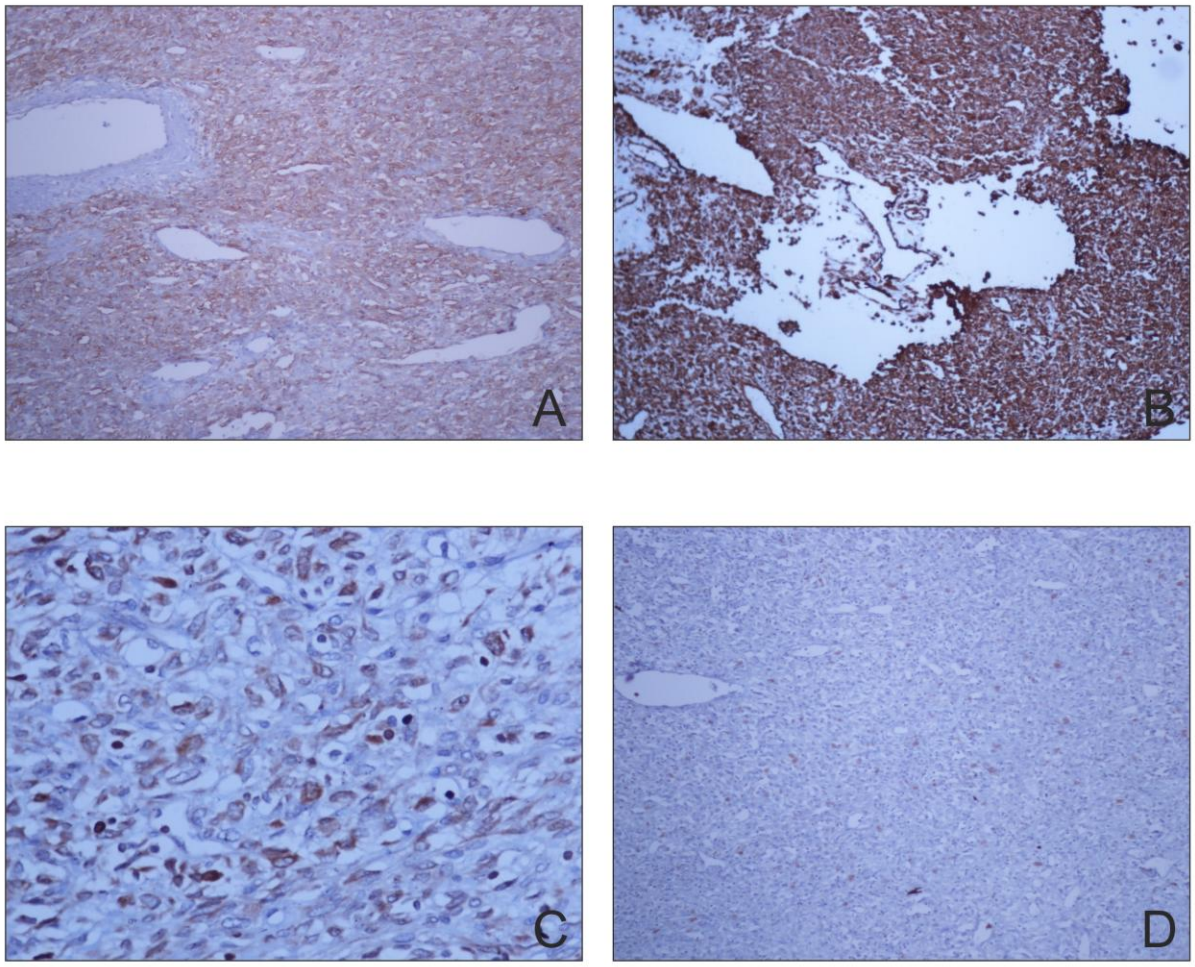

Figure 2: Immunohistochemistry. (A) Diffuse positive CD 34 staining (40x). (B) strong diffuse staining with CD 99 (10x). (C) Tumor cells showing positivity with bcl 2 (40x). (D) Tumor cells are negative for S100 (4x).

DISCUSSION

Lipomatous hemangiopericytoma is a rare variant of hemangiopericytoma, which occurs in middle-aged adult patients (21-79 years) and presents clinically as a long standing, deep seated and indolent soft tissue tumor. These tumors have a wide anatomical distribution including the lower extremities, head & neck region, mediastinum and pelvic cavity but occur most frequently in the thigh, orbit and retroperitoneum. A case of LHPC has also been reported in the stomach. [3] These tumors behave in a benign fashion, and are not known to recur.

Grossly, LHPC presents generally as a well-demarcated, variably encapsulated, medium sized tumor. Cut section shows a lobulated surface with alternating areas of tan white to yellowish tumor tissue.

Microscopically, it is characterized by a varying combination of cellular areas composed of round to spindle cells, collagenous or myxoid stroma with focal sclerotic areas; hemangiopericytoma like vasculature made of medium to small-sized thick-walled branched vessels, and lipomatous areas composed of mature adipocytes. [3] Whether the presence of fat in LHPC results from a nonspecific entrapment phenomenon or, instead, represents a specific line of differentiation remains unsolved. [4] Because these tumors appear encapsulated, and the lipomatous changes are present throughout the tumor, suggests that fat is an integral component of the tumor and non-specific peripheral adipose tissue entrapment appears unlikely. Moreover LHPC has been reported in locations where fat does not occur (sinonasal passages). [5]

Hemangiopericytoma like vascular pattern may occur in many soft tissue tumors, including synovial sarcoma, mesenchymal chondrosarcoma and benign fibrous histiocytoma. Due to a lack of specificity of light microscopic appearance, ultrastructural characteristics and immunohistochemical staining profile of hemangiopericytomas many authors reserve this term for soft tissue tumors that lack evidence of alternative lines of differentiation. [6]

The recent WHO classification has suggested a close relationship between hemangiopericytoma and solitary fibrous tumors. However, differences between the two lesions exist. Vascular prominence always forms a prominent component of the architecture of hemangiopericytomas when viewed under low power. This is in contrast to solitary fibrous tumors, in which architectural variability is a hallmark. Guillou et al proposed that as the morphological, immunohistochemical and ultrastructural features of LHPC and solitary fibrous tumor overlap, in all likelihood LHPC represents a fat containing variant of solitary fibrous tumor.

Lipomatous hemangiopericytomas should be distinguished from all those tumors showing a dual adipocytic and spindle cell composition. Liposarcoma, spindle cell lipoma, myolipoma and agiomyolipoma are important differential diagnostic considerations. LHPC lacks pleomorphism, significant mitotic activity, sclerotic zones with atypical cells and lipoblasts, thus allowing their distinction from liposarcomas. Myxoid liposarcomas display a prominent vascular pattern and the vascular configuration is an important clue in distinguishing between liposarcoma and LHPC. Myxoid liposarcomas demonstrate a delicate plexiform capillary vascular network, unlike the staghorn pattern of LHPC. Occasionally some well differentiated liposarcomas maybe admixed with a low grade spindle cell component. However, these usually show a fascicular leiomyosarcoma like growth pattern and shows immunohistochemical reactivity with smooth muscle markers. [4] These lesions are ordinarily easily distinguished in entire resection specimens, but can be problematic in context of limited small biopsies.

Spindle cell lipoma almost exclusively presents in the subcutaneous tissues of the head and neck. It contains bland spindle cells and bundles of wiry collagen. Because of strong morphologic resemblance, myolipoma is likely to be confused with LHPC. However spindle cells in the former lesion are arranged in a fascicular pattern and show diffuse positive staining for SMA and desmin. [4] Angiomyolipoma can be distinguished due

to its common relationship to the kidney, distinctive arrangement of spindle cells around thick walled vessels and their positivity with smooth muscle markers, HMB45 and Melan A.

Immunohistochemistry shows the non-adipocytic component of LHPC to be positive for CD34, CD99 and bcl-2 as opposed to other above-mentioned lesions.

In summary, lipomatous hemangiopericytoma is a rare variant of hemangiopericytoma, which behaves, in a benign fashion. An important implication is its distinction from liposarcoma, which can be particularly difficult when dealing with biopsy material.

REFERENCES

1. Nielson GP, Dickersin GR, Provenzal JM, et al: lipomatous hemangiopericytoma. A histologic, ultrastructural and immunohistochemical study of a unique variant of hemangiopericytoma. Am J Surg Pathol 1995;19:748-756.
2. Guillou L, Fletcher JA, Fletcher CDM. Extrapleural solitary fibrous tumor and hemangiopericytoma. In: Fletcher CDM, Unni KK, Mertens F, editors. World Health organization classification of tumours: pathology and genetics of tumors of soft tissue and bone. Lyon: IARC Press, 2002; 86-90.
3. Jing HB, Meng QD, Tai YH. Lipomatous hemangiopericytoma of the stomach: A case report and a review of literature. *World J Gastroenterol* 2011;17(43): 4835-4838.
4. Guillou L, Gebhard S, Coindre JM. Lipomatous Hemangiopericytoma: A fat containing variant of solitary fibrous tumour? Clinicopathologic, immunohistochemical and ultrastructural analysis of a series in favor of a unifying concept. Hum Pathol 2000;31:1108-1115.
5. Folpe AL, Devaney K, Weiss SW. Lipomatous hemangiopericytoma: A rare variant of hemangiopericytoma that maybe confused with liposarocma. Am J Surg Pathol 1999; 23(10): 1201-1207

6. Ceballos KM, Munk PL, Masri BA, O'connell JX. Lipomatous hemangiopericytoma- A morphologically distinct soft tissue tumor. Arch Pathol Lab Med 1999;123:941-945.