

# Case Report

## Benign retroperitoneal mesenchymoma of childhood

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### ABSTRACT

Benign mesenchymoma is a rare tumor composed of mature fat, thin small and medium-sized vessels and spindle cells of smooth muscle origin and has been found in variable locations. There have been only three reports of such cases in the retroperitoneal location. We report one such rare retroperitoneal tumor.

**KEY WORDS:** Benign mesenchymoma, lipomatous tumor, retroperitoneal tumor

### INTRODUCTION

Benign mesenchymoma is a rare tumor composed of an admixture of two or more mature mesenchymal tissue types found in variable locations such as the mesentery, skin, liver and ovary.<sup>[1]</sup> Mesenchymomas are clinically and histologically benign. Only three retroperitoneal cases have been reported so far in literature.<sup>[1-3]</sup> This is a report of one such rare retroperitoneal tumor along with a review of other reported retroperitoneal lipomatous tumors of childhood.

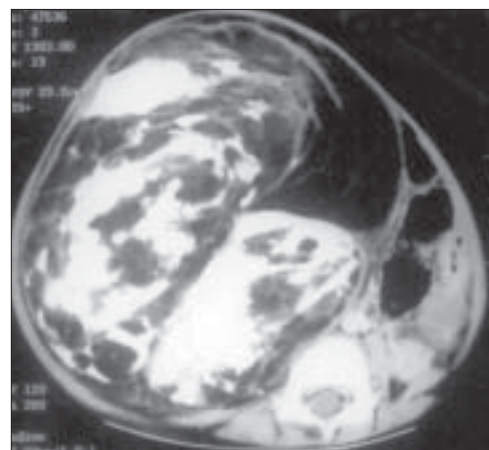
### CASE REPORT

A five year-old female presented with a huge, painless, gradually enlarging intraabdominal lump of three years' duration. The child had no history of vomiting, bowel complaints, weight loss, hematuria or urinary disturbances. The reason for the hospital visit was discomfort due to weight of the tumor. On examination, the child had mild pallor although the respiratory and cardiovascular systems were normal. A large, nontender; immobile, firm mass with a smooth surface and well-defined margins occupied the entire abdomen. There was no fluid thrill or bruit. On per rectal examination, a hard mass was palpable anteriorly. Hematological parameters, renal function tests and liver function tests were normal.

Plain X-ray of the abdomen showed a large calcified mass, predominantly occupying the right half of the abdomen crossing the midline and pushing the bowel towards the left side. Ultrasonography revealed a large heterogenous calcified mass displacing the normal liver, spleen, kidneys and the urinary bladder. The kidneys could be seen being

separate from the mass. Computed tomography (CT) revealed a large solid calcified retroperitoneal mass displacing the normal viscera. The aorta and inferior vena cava (IVC) were displaced towards the left side and the right iliac vein was stretched over the tumor [Figure 1]. The child underwent exploratory laparotomy revealing a large retroperitoneal tumor 30 x 30 cm occupying the entire abdomen and pushing the bowel towards the left side. The right kidney was compressed by the tumor. Although the aorta, IVC and iliac vessels were pushed and stretched by the tumor, they were not invaded by it. The tumor was adherent to the soft tissues in the right iliac fossa, well-encapsulated and could be removed *in toto*. The cut surface of the tumor revealed fat and extensive bony areas that were very hard to cut.

Histological examination showed that the tumor was mainly composed of mature fat tissue divided into



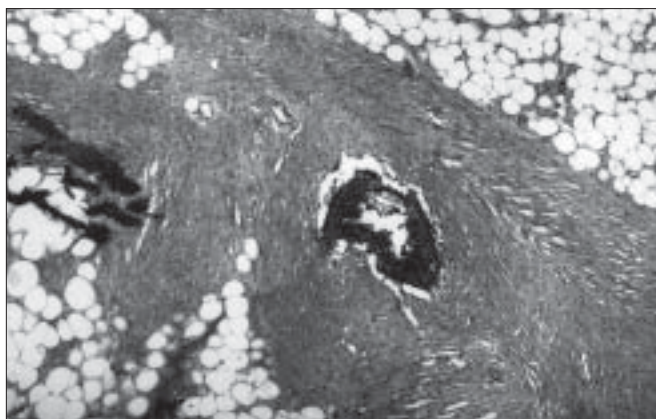
**Figure 1:** Computed tomography showing a large solid calcified retroperitoneal mass displacing the normal viscera, aorta and IVC

lobules by fibrocollagenous bands. In addition, the tumor had bone, cartilage, blood vessels and spindle cells of smooth muscle origin suggestive of a benign mesenchymoma [Figure 2]. The lymph nodes had features of reactive hyperplasia.

## DISCUSSION

Benign mesenchymoma are rare tumors. Stout described these tumors to be of mesenchymal origin that differentiate into a varied assortment of tissues.<sup>[1]</sup> Diagnosis is made when at least two distinct mesenchymal elements are present, which are not ordinarily found in a tumor, like bone and cartilage. Since the fibrous element is present in all the mesenchymal tumors, this must not be counted as one of the features for diagnosis. The tissues in benign mesenchymoma include fat, blood vessels, smooth muscles, striated muscle, cartilage, lymphoid and hematopoietic tissue.<sup>[1]</sup> A negative S-100 can exclude spindle cell lipoma or Schwann cell tumor.<sup>[2]</sup> Stout stated that the tumor occurred in both benign and malignant forms and that while the benign form was capable of local recurrences, the malignant form may have distant metastasis.

These tumors might be called 'Hamartomas'. However, such types of tumors are found both in skin and renal areas as well, which have epithelial elements that do not enter into the composition of the tumor. In Le Ber and Stout's opinion, this tumor should be called Benign Mesenchymoma.<sup>[3]</sup> They reported 39 cases of benign mesenchymomas in children. Since then, additional reports have appeared such as benign mesenchymoma in the sole of a 7 year-old child,<sup>[4]</sup> in the left femoral triangle in a nine month-old male,<sup>[5]</sup> 38 cases of chest wall benign mesenchymoma in children,<sup>[6]</sup> 36 cases of head and neck mesenchymoma including 15 cases arising from tongue, out of which six presented in childhood, and<sup>[7]</sup> benign renal mesenchymoma in a 13 year-old girl.<sup>[8]</sup>



**Figure 2:** Photomicrograph showing islands of mature fat tissue, bone formation, blood vessels and bundles of spindle cells (H/E, x120)

A thorough search of literature revealed only three cases of retroperitoneal benign mesenchymoma—ours being the fourth.<sup>[3,9]</sup>

If a large retroperitoneal soft tissue mass is present with several sarcomatous features such as prominent calcification / ossification or a fatty component, the differential diagnosis should include a malignant mesenchymoma.<sup>[10]</sup> However, pleomorphic and hyperchromatic blast cells suggestive of malignancy were not identified in our case.

The most common differential diagnosis of a retroperitoneal lipomatous tumor is angiomyolipoma that usually arises from the kidney. Hruban *et al.* emphasized the need to include this rare tumor in differential diagnosis for well-differentiated fatty tumors occurring outside the kidney.<sup>[11]</sup>

Hence, the differential diagnosis of a well-differentiated fatty neoplasm should include spindle cell lipoma, atypical lipoma, liposarcoma, malignant mesenchymoma and angiolipoma. Spindle cell lipoma and atypical lipoma in the retroperitoneum also may behave in a malignant manner and are best considered as well-differentiated liposarcomas.<sup>[12]</sup>

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