

SCHWANNOMA IN RETROPERITONEUM – A FREQUENT TUMOR AT AN IN-FREQUENT LOCATION.

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Abstract: Schwannomas are benign tumors originating from Schwann cells of peripheral and cranial nerves mostly occurring in female patients 20-50 years of age. It is rare to find schwannoma in retroperitoneum. They are difficult to diagnose at this site unless they cause compression symptoms. They are also sometimes diagnosed incidentally during surgery. We here present a case of a 59 year old female who presented with 5 year history of epigastric pain which was associated with post prandial vomiting and constipation. A retroperitoneal cystic mass in the vicinity of right adrenal gland was found on radiology. Excision of the mass was done and sent for histopathology in the Department of Pathology, Lahore Medical & Dental College, Lahore Pakistan. The received specimen comprised of a fragmented cystic biopsy. Histological examination revealed a spindle cell neoplasm comprising round to spindle cells arranged in hypo cellular and hyper cellular areas with negligible mitotic activity. Immunohistochemical analysis revealed strong positive S100 protein in the spindle cells which is a distinct immunohistochemical marker for schwannoma. Schwannoma in retroperitoneum is a rare occurrence. It is important to differentiate it from the malignant counterparts which are more common in this location and require more extensive surgery and treatment.

Index Terms: Schwannoma, retroperitoneum, infrequent, location

I. INTRODUCTION:

Schwannomas are benign soft tissue tumors twice common in females than in males with the age range of 20–50 years [1]. The origin of this tumor is Schwann cells of the peripheral nerve sheath. They can arise at places where the Schwann cells are located: e.g. nervous system, trunks and extremities. Retroperitoneal schwannoma is an uncommon diagnosis. It constitutes only 0.5%–5% of all schwannomas. Commonly encountered tumors in retroperitoneum are malignant rather than benign. [2] Retroperitoneal schwannomas (RS) present as an incidental finding and are diagnosed when they produce compression symptoms due to their large size. These tumors have a greater tendency to undergo sudden degenerative changes and/or cause hemorrhage in this deep location. [1]

II. CASE PRESENTATION:

We report a case of 59 year old female named MB who presented with 5 year history of intermittent and colicky epigastric pain radiating to right hypochondrium and associated with post prandial vomiting. The patient also had history of constipation. On abdominal ultrasound, a retroperitoneal cystic mass was found in the vicinity of right adrenal gland. The mass was excised and sent for histopathology in the Department of Pathology, Lahore Medical & Dental College, Lahore. Gross examination revealed a fragmented cystic biopsy comprising a larger fragment measuring 5x5x3cm and multiple small fragments measuring 4x2x1cm in aggregate as shown in Fig.1.



Figure.1 Gross picture showing cystic fragments

Histological examination revealed cystic and solid areas. Cystic areas showed proliferation of spindle cells lined by foamy and hemosiderin laden macrophages as evident from Fig.2. Solid areas were hyper cellular showing proliferation of spindle cells with dilated and hyalinized blood vessels as depicted in Fig.3 and Fig.4 respectively. No atypia, mitosis or areas of necrosis were seen. On the basis of available clinical, radiological data and microscopic examination, list of differentials included gastrointestinal stromal tumor, Leiomyosarcoma, Hemangioma and Schwannoma. To rule out all the possible differentials, the specimen was extensively sectioned but the morphology remained the same. The slides were subjected to immunohistochemical analysis which revealed strong positive S100 staining of the spindle cells with negative Desmin and DOG-1 staining as shown in Fig.5. Based on histopathology and immunohistochemistry, diagnosis of spindle cell neoplasm favoring Schwannoma was made.

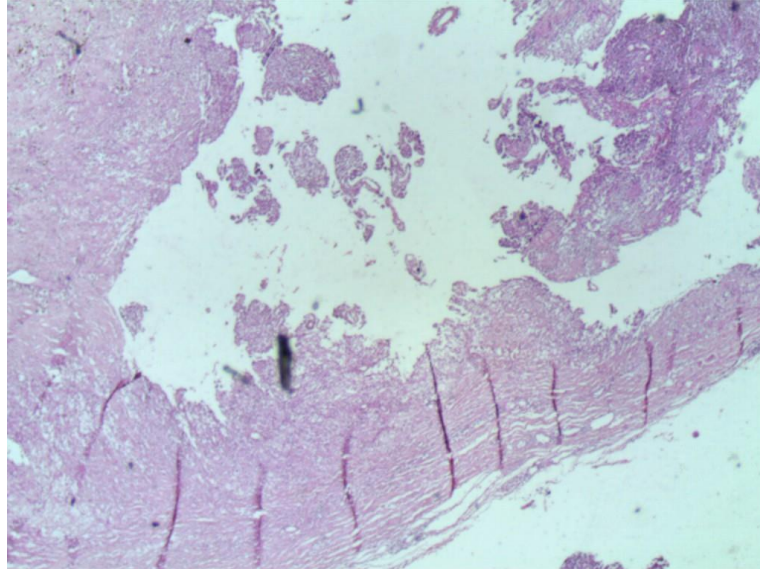


Figure.2 Cystic areas on microscopy

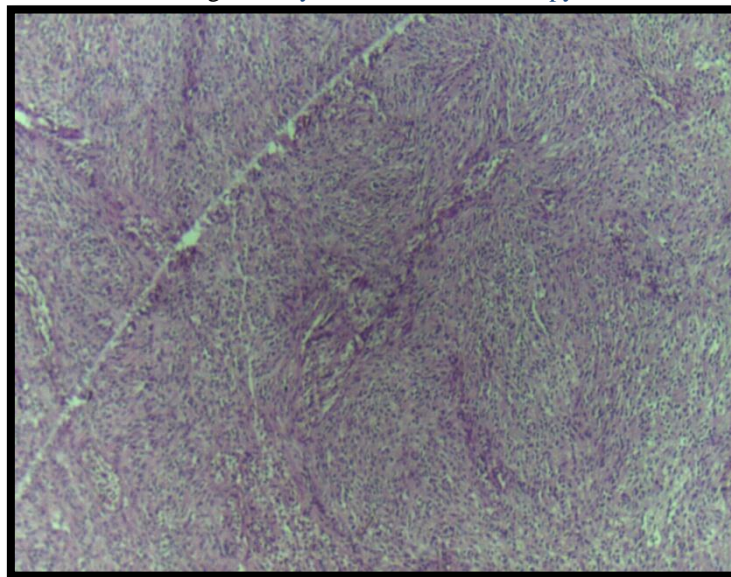


Figure.3 Hyper cellular areas showing proliferation of spindle cells

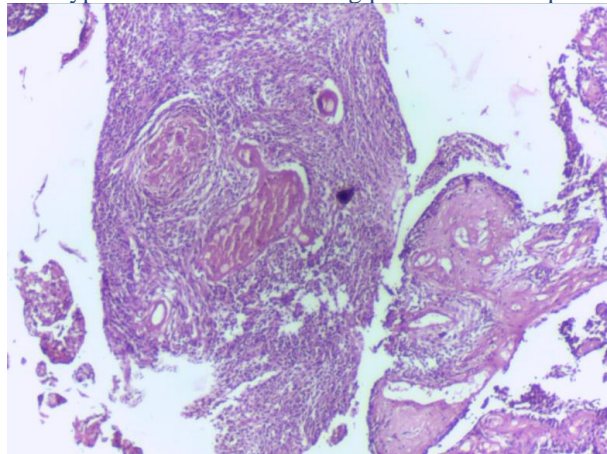


Figure.4 Dilated and hyalinized blood vessels

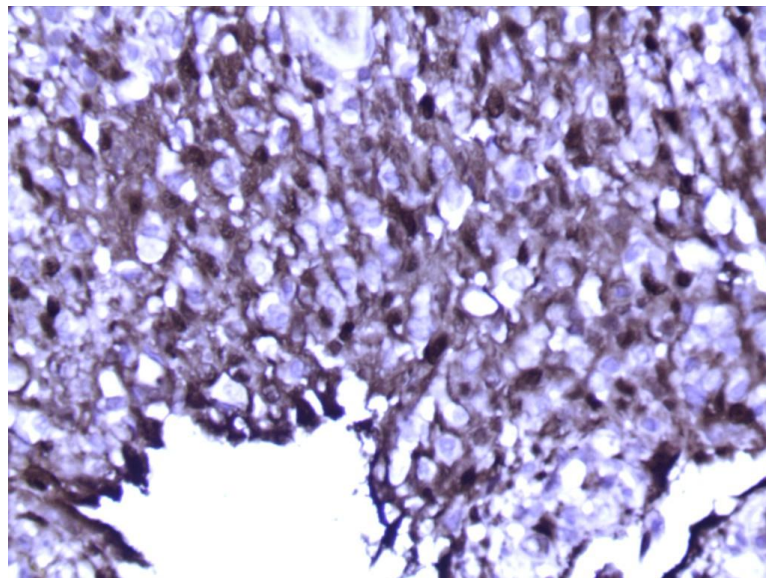


Figure.5 Positive S100 IHC stain

III. CASE DISCUSSION:

Schwannomas are tumors of neuroectodermal origin that are usually solitary, noninvasive and rarely show malignant transformation unless there is a syndromic association. [3] These tumors are generally encapsulated and originate from the Schwann cells of the peripheral or cranial nerve sheaths. Clinically, the patients present with non-specific symptoms which depend on the location and size of the lesion. [4] Schwannomas are usually encountered in young to middle-aged adults with a female predominance. [5]

Retroperitoneum is a rare site for schwannoma to occur. They usually escape detection and reach a huge size (average 9cm in diameter) due to the large capacity of the retroperitoneum to accommodate a growing mass. Retroperitoneal schwannomas are diagnosed either on radiology as an incidental finding or per-operative at time of surgery. [6] Clinical presentation is late when patient becomes symptomatic due to large size. [7] The compression symptoms include vague abdominal pain, abdominal mass and sometimes urinary difficulties [3] They can also cause abdominal distension, pain in lower back, sensory and motor disturbances along with pain in the lower extremities[8].In this case there was a five years long history of epigastric pain, intermittent and colicky in nature, radiating to right hypochondrium. It was associated with post prandial vomiting and constipation.

The characteristic histological features of schwannoma are identification of Antoni A and Antoni B areas. Antoni A areas are hyper cellular and are composed of spindle cells arranged in short bundles or fascicles leading to Verocay formation, while Antoni B areas are hypo cellular and can show myxoid and cystic degeneration.[1]In our case foamy and hemosiderin laden macrophages along with hyalinized blood vessels were also evident.

There is a long list of differential diagnoses for a mass in retroperitoneum, which includes paragangliomas, pheochromocytomas, and histiocytic sarcomas. Correlation with radiological and anatomopathological examination is mandatory to rule out [4] malignant sarcomas which are more common in the retroperitoneum than their benign counterparts. [2]

In this case study; taking account of provided clinical information, gross and microscopic examination, initial diagnosis of spindle cell neoplasm with the differential diagnosis of Schwannoma, GIST, giant Liposarcoma, Leiomyosarcoma and hemangioma was made. The diagnosis was confirmed by immunohistochemical analysis, which showed strong diffuse positivity of the spindle cells for S100 protein. A strong diffuse positivity of the spindle cells for S100 protein is a salient immunohistochemical property of schwannoma [9]

Complete surgical excision by either open or laparoscopic procedure is considered the treatment of choice for schwannomas; because their sensitivity to radiotherapy and chemotherapy is not reported. [10] Local recurrence has been documented in case of incomplete resection. [5]

IV. CONCLUSION:

A case of schwannoma in retroperitoneum is being reported because it's a rare and incidental finding at this site in a middle to old age female. Surgeons were suspecting it to be a GIST or a giant sarcoma but histopathological examination revealed a spindle cell neoplasm favoring schwannoma which was confirmed with the help of positive S100 immunostain. It is important not to over diagnose it as a malignant sarcoma because they are more common in the retroperitoneum and need extensive surgery and treatment while schwannoma is simply treated by complete excision.

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