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Case Report

Mesenteric schwannoma: a rarity or rising trend?

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ABSTRACT

Schwannomas are neurogenic, benign, well circumscribed tumors arising from schwann cells. Schwann cells are neural originating cell. These are either sporadic or inherited with sporadic being most common. In about 3% of patients, association with neuro-fibromatosis 2 gene is documented. Primary mesenteric schwannoma is extremely rare with establishing a pre-operative diagnosis almost impossible. Imaging modalities including computed tomography as well as MRI fails to make a diagnosis due to the non-specific characteristics. Surgery is curative with histopathological examination and immunostaining establishing the final diagnosis post-operatively. Immunostaining helps in differentiating it from other conditions such as gastro-intestinal stromal tumors as well as malignancies. Rarity of this condition make this case report of a 32-year-old male unique.

Keywords: Schwannoma, Benign, Mesenteric, Laparotomy, S-100

INTRODUCTION

Schwannomas are benign tumors that exhibit Schwann cell differentiation and often arise directly from Schwann cells of peripheral nerves.[1] Usually solitary sporadic lesions, both sporadic as well as inherited schwannomas are commonly associated with inactivating mutations in the neurofibromatosis2 (NF2) gene on chromosome 22 which encodes for the protein Merlin.[2] Loss of expression of the NF2 gene product, Merlin, is a consistent finding in all schwannomas. The exact mechanism of tumorigenesis and role of Merlin is poorly understood.[3] Schwannomas are usually found in head, neck, extremities, mediastinum and retroperitoneum. Intrabdominal schwannoma are very rare and occurs most commonly in alimentary tract.[4] Primary mesenteric schwannoma is extremely rare. In this case report, we present a rare case of mesenteric mass in a young individual.

CASE REPORT

A 32-year-old male, resident of Uttar Pradesh, India presented to a peripheral center with complaints of intermittent right lower abdominal pain of one-month duration in February 2018. On initial evaluation, he was detected to have a vesico-ureteric junction calculus and was referred to our center for further management. On admission patient reported history of recurrent pain over the right flank which was colicky in nature and non-radiating. However, there was no history of nausea, vomiting, constipation, obstipation or any urinary complaints. His vital parameters were within normal limits. Physical examination of the abdomen revealed mild tenderness in right flank with no palpable lump or signs of bowel obstruction.

Initial sonography of whole abdomen showed right vesico-ureteric calculus 4×3 mm and a soft tissue mass in left para-aortic region. On detailed imaging evaluation

using computed tomography (CT) with contrast, a 5.3 mm calculus was seen in vesico-ureteric junction and a well-defined heterogeneous lesion of 35.3×42.4×36.6 mm was seen along small bowel mesentery, anterior to aorta just above aortic bifurcation at L3 vertebrae with minimal enhancement. Few scattered spicules of calcification were seen with no obvious fat stranding. A radiological diagnosis of suspected desmoid tumor of small bowel mesentery was made.

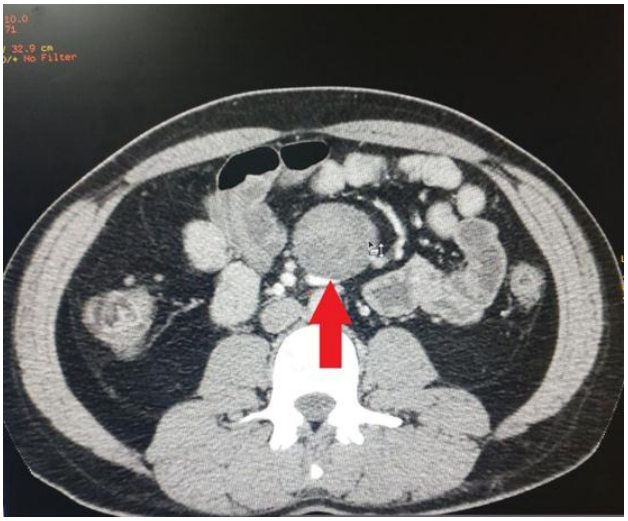

Figure 1: CECT abdomen-well encapsulated mass in mesentery in lower abdomen (red arrow).

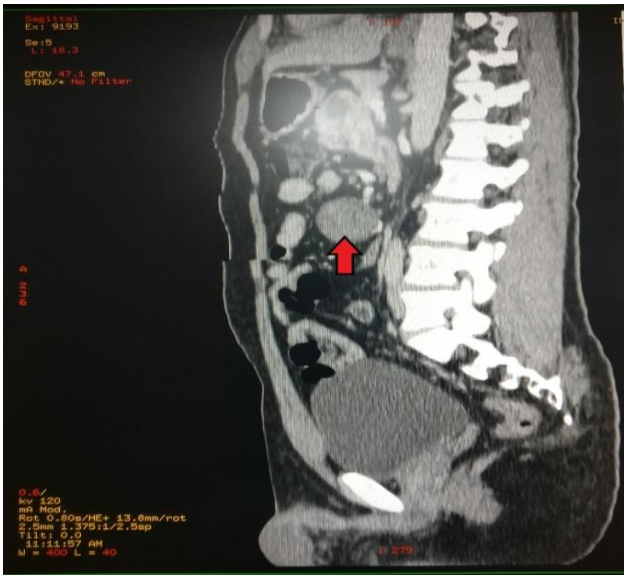

Figure 2: CECT abdomen (sagittal view): mass arising from mesentery not abutting any major structures.

Patient underwent exploratory laparotomy under general anaesthesia. Intraoperatively a 5×4 cm mass was found distal to Duodeno-Jejunal flexure in midline over jejunal mesentery. This was a well encapsulated mass lying over a blood vessel. Wide local excision was done for the mass along with resection anastomosis of Meckel's diverticulum and appendectomy. Drain was placed in pelvis and removed on post-operative day three. Post op recovery of the patient was uneventful. He was discharged on post op day eight. He was further referred to Urologist for the vesico-ureteric calculus. Patient has been on regular follow up since with no evidence of recurrence till date.

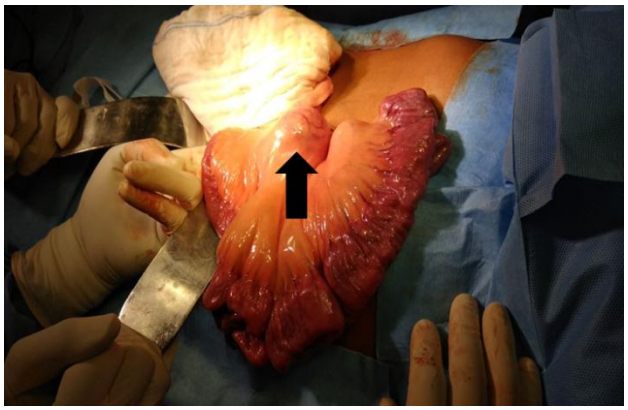

Figure 3: Per operative image- mass arising from jejunal mesentery, well encapsulated lying over blood vessel (black arrow).

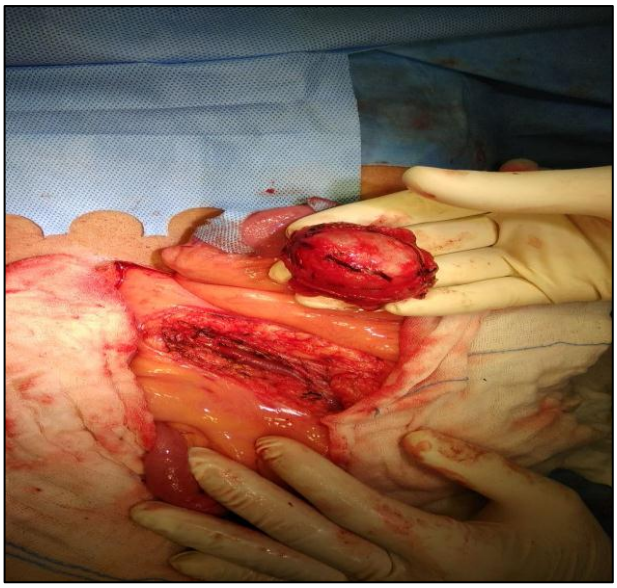

Figure 4: Per operative image showing completely excised mass from jejunal mesentery shaven of the underlying blood vessels.

On gross examination, the tumor was a hard-round mass. Histopathological examination revealed both hyper and hypocellular areas. Immunohistochemistry (IHC) was strongly positive for S-100, while SMA, CD-34, CD-117 were negative. Ki-67 was less than 2%.

DISCUSSION

Schwannomas are well circumscribed, encapsulated, benign (90%) masses that abut the associated nerve

without invading it.[5] It usually occurs in young to middle aged patients, with equal sex distribution.[4] Schwannoma is usually seen in head, neck and extremities.[6] Tumors arising in ligament, mesentery and intra-abdominal organs are extremely rare.[5,7-10] Secondary degenerative changes like cyst formation, hemorrhage, calcification, hyalinization and rarely ossified degeneration are seen sometimes.[3]

Mesenteric schwannoma is extremely rare entity with approximately 10 documented cases till 2018.[3,8,11-18] These are usually detected incidentally or with vague nonspecific abdominal symptoms such as chronic pain. It is very difficult to detect preoperatively due to lack of specific radiological characteristics. The final diagnosis is based on histopathological examination and immune-histochemistry.[12] Ultrasonography usually show well defined hypo-dense lesions.[15] Computed tomography (CT) scans reveal well circumscribed, round-oval, homogeneous masses. On contrast-enhancement, they appear as a heterogeneous mass. MRI findings are again non-specific and only reveal low-intensity images on T1-weighted imaging and high-intensity images on T2-weighted imaging, due to Antoni A and Antoni B areas and secondary degenerative changes. However, these imaging modalities only help us to determine the size, location and extension only and will not aid in making a definitive diagnosis.[19]

Grossly these tumors are firm, grey masses. Microscopically, they are comprised of admixture of dense and loose areas referred respectively to as Anton A (closely packed spindle cells) and Anton B areas (hypocellular with myxoid tissue and more water content).[20,21] Schwann cells are characterized by presence of spindle elongated nucleus with a wavy or buckled shape. Schwannoma which lack Anton B areas may resemble sarcoma. Malignant transformation is very rare.[18] Schwannomas show strong immunoreactivity for S-100 protein, while CD34, CD117, DOG-1 and SMA are negative similar to the IHC of the present case.[22] Ki-67 labelling indices will help as distinguish between problematic schwannomas and malignant peripheral nerve sheath tumors. Indices > 20% are highly predictive of malignancy.[23]

Surgical intervention with wide local excision (WLE) of the schwannoma is curative with very low recurrence rates.(3) In our case post op follow up was done post WLE for 18 months with no evidence of any recurrence.

CONCLUSION

Mesenteric schwannomas are very rare tumors, with approximately 10 cases reported till date worldwide. Pre-operative diagnosis is almost impossible due to non-specific symptoms and imaging characteristics. Histopathological examination combined with immunostaining will help in establishing diagnosis as well as differentiating it from other benign as well as malignant conditions. Surgery is curative as these are mostly benign. A differential diagnosis of Schwannoma can be kept in mind while addressing such patients who are not fitting into the common diagnosis clinically or radiologically.

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