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Incidental Retroperitoneal Ancient Schwannoma.

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ABSTRACT

Ancient schwannomas are degenerate peripheral nerve sheath tumors that very rarely occur in the retro peritoneum. They generally reach large proportions before producing symptoms usually only due to mass effect. Here, we describe an incidentally detected retroperitoneal schwannoma in an abdominal computerized tomography (CT). A 42 year old male presented with complaints of fever and lower abdominal pain for 48 hours was incidentally found to have a mass in retro peritoneum in the left para aortic region. The patient successfully had excision of the tumor. Histological examination showed encapsulated tumors that displayed alternating areas of dense cellularity and areas of myxoid matrix consistent with a diagnosis of ancient schwannoma. A diagnosis of ancient schwannoma should be entertained for any heterogeneous, well encapsulated mass in the retro peritoneum. In these cases less radical surgical resection should be considered as malignant transformation of these tumors is extremely rare and recurrence is uncommon following excision. Rare lesions with benign course such as schwannoma can be detected incidentally.

Keywords: Retroperitoneal, Schwannoma, abdominal pain.

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CASE REPORT

A 42 year old male presented with complaints of lower abdomen pain and fever of two days duration. The pain was intermittent, dull aching with no specific history of radiation or migration. He had fever for 2 days prior to presentation with nausea and vomiting. He also gave a history of dysuria. On examination he had tenderness in the right hypogastric region. His total leucocyte and neutrophil counts were elevated. Urine routine examination was normal. Other blood investigations were normal. He was subjected to an ultrasound of the abdomen which revealed well defined hypoechoic lesion in left para aortic region. The report suggested the possibility of the lesion being a lymph node / para ganglioma. A contrast enhanced CT scan of the abdomen was done which showed a well-defined moderate sized heterogeneously enhancing mass lesion in the left para aortic region at L2, L3 level. No evidence of calcifications within the mass lesion or encasement of adjacent aorta was found. There was no evidence of any intraspinal extension of lesion. CT scan reported the possibility of the lesion being a retroperitoneal tumour / lymph nodal mass. CT guided FNA (Fine Needle Aspiration) was done and reported as a retroperitoneal sarcoma [1,2].

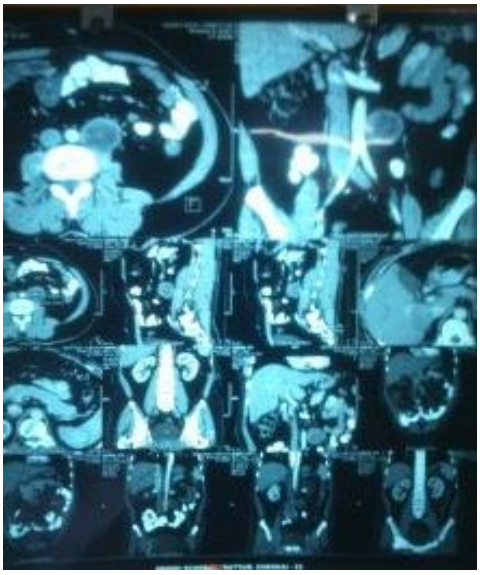

Figure 1: The CT Image of the lesion

A laparotomy was planned and done. A midline incision made and the abdomen opened. The left paracolic cutter mobilized and swelling measuring about 5 x 5cm noted between aorta and left ureter. Meticulous dissection was done and the feeding vessels were ligated. Swelling was excised in toto.

Figure 2: The retroperitoneal mass

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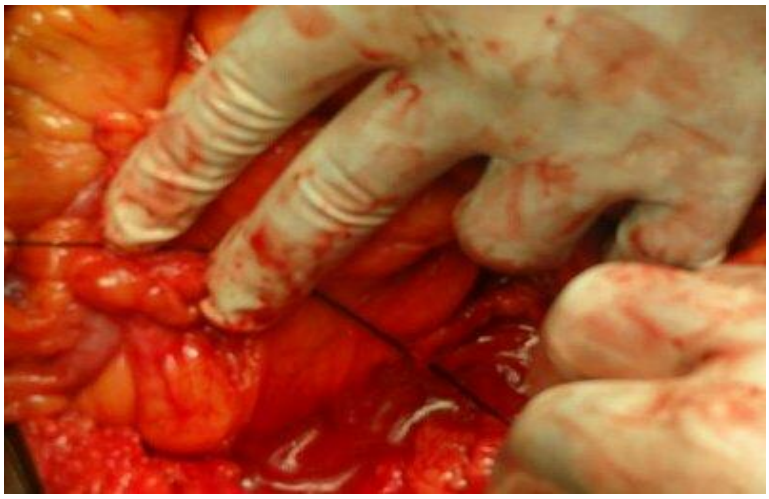

Fig 3: Ligating feeder vessels

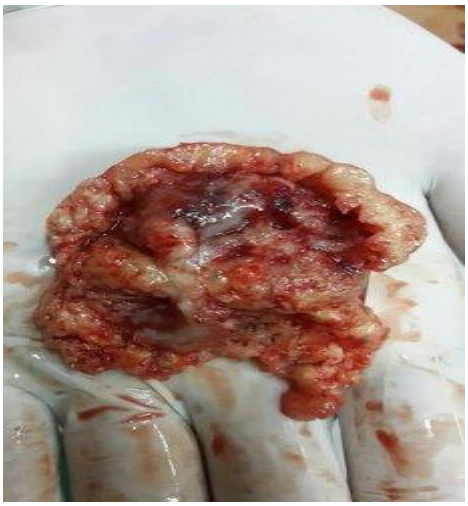

Figure 4: Cut section of the specimen

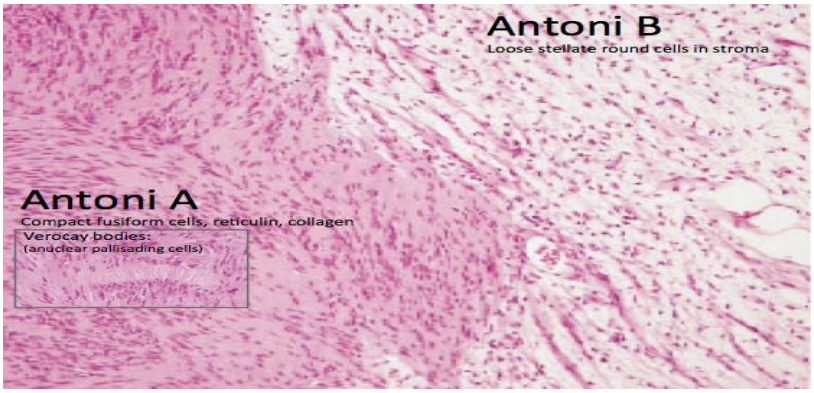

Figure 5: Histopathological picture of the specimen

DISCUSSION

Schwannomas (neurilemmomas) are benign soft tissue neurogenic tumors that arise from Schwann cells of peripheral nerve sheaths. They usually arise from sensory nerves. They most commonly manifest in the head and neck region and in the extremities. Retroperitoneal schwannomas are rare and account for 0.7% to 2.7% of these tumors [3].

Ancient schwannomas are a rare variant of schwannomas, originally described by Ackerman and Taylor in 1951 [4]. They account for 0.8% of soft tissue tumors. They are characterized by distinctive degenerative tumor features including cystic necrosis, stromal edema, xanthomatous change, fibrosis, perivascular hyalinization, calcification and degenerative nuclei with pleomorphism, lobulation and hyperchromasia [4,5]. These degenerative features are attributed to the growth and "aging" of the tumor, hence the term "Ancient schwannoma." Growth of the tumor over time leads to vascular insufficiency, with resulting areas of tumor degeneration. Previous studies have correlated tumor size with progressive degenerative features [6]. Despite these degenerative changes, ancient schwannomas behave similar to their conventional counterparts. They are benign, slow-growing tumors with rare malignant transformation [7].

On histology, ancient schwannomas shows areas of cellularity and areas of myxoid matrix, as also observed in conventional schwannomas. There is, however, a relative loss of cellular regions, which tend to be fibrosed or sclerotic. These areas may degenerate into hematomas and cysts, leading to an overall decreased density. Nuclear palisades, seen in classic schwannomas, are absent and large intra-nuclear invaginations are characteristically present [8]. The degenerative histological features of ancient schwannomas are evident in their radiographic features as well-circumscribed complex cystic masses with inhomogeneous contrast enhancement as noted in the cases presented. Non-enhancing areas on CT imaging correspond to regions of cystic degeneration, with contrast enhancement seen in surrounding tissues [9]. MRI with gadolinium enhancement has been advocated as superior to CT in demonstrating tumor cystic degeneration, defining margins and in some cases identifying the point of neuronal origin [9,10]. However, radiographic modalities do not differentiate benign from malignant disease unless tumor invasion or metastasis is seen. Increased accumulation of 2-deoxy-[(18)F] fluoro-D-glucose (FDG) on PET scanning has been previously reported in cases of schwannomas, and was noted in one case in our series [11]. The role of PET in assessing the malignant potential of schwannomas is however undetermined. Surgery is usually required for definitive diagnosis of these tumors and differentiation from other retroperitoneal malignancies [12].

Tumor enucleation, with preservation of vital structures in the vicinity, is the preferred surgical approach when a diagnosis of retroperitoneal schwannoma is highly suspected, since these tumors have not been reported to recur following excision [13].

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