

Retroperitoneal Schwannoma: A Case Report

Dr. Dhruv Gupta¹ Dr. Deepika Sharma² Dr. KS Mehta³ Dr. Gurpreet⁴ Dr. Amit Suri⁵

1. Postgraduate Student Department Of Surgery ASCOMS And Hospitals Jammu (J&K) India

2. Postgraduate Student Department Of Surgery ASCOMS And Hospitals Jammu (J&K) India.

3. Professor Department Of Surgery ASCOMS And Hospitals Jammu (J&K) India.

4. Assistant Professor Department Of Surgery ASCOMS And Hospitals Jammu (J&K) India.

5. Associate Professor (Urology) Department Of Surgery ASCOMS And Hospitals Jammu (J&K) India.

Abstract: Retroperitoneal schwannoma is a very rare primary neurogenic tumour of the retroperitoneum with limited reporting of cases in the literature. It usually affects adults aged 20 to 50 years and has a male predominance. Symptomatology of schwannomas is highly nonspecific and depends on the location and size of the lesion. The majority of retroperitoneal schwannomas are benign in nature although malignant ones have also been reported. Its diagnosis and management poses a challenge for the radiologist and the surgeon. Authors are describing a case of "RETROPERITONEAL SCHWANNOMA" which was reported in their centre.

Keywords: Retroperitoneal, Schwannoma, Neurogenic, Nonspecific, Challenge.

I. Introduction:

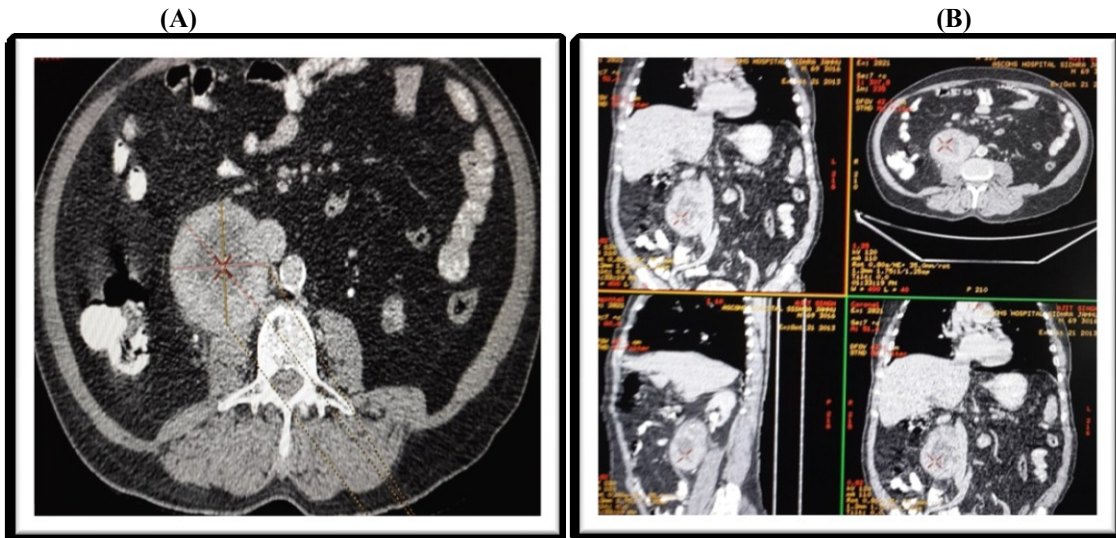
Schwannomas are slow growing solitary neural sheath tumors that originate from schwann cells. Commonly arising from cranial and peripheral nerves in the head and neck region or in the upper extremities [1]. However schwannomas may appear in the posterior mediastinum but rarely in the retroperitoneum, comprising 0.3% to 3.2% of all schwannomas [2]. In retroperitoneum they can grow to a large size, sometimes displacing, and more rarely invading surrounding structures and organs before becoming clinically apparent. Presentation of the patient is often varied and sometimes its an incidental finding. Radiology can be used to make the diagnosis, to assess the extent of the lesion, to look for features of malignant transformation but a correct preoperative diagnosis is difficult to make and these tumors are often misdiagnosed. Tumour excision in toto is considered the treatment of choice. Confirmation of diagnosis is based on post operative histopathologic examination demonstrating specific Antoni A/B areas and characteristic immunohistochemical features i.e. positive S-100 and a negative CD-34 [3]. The prognosis of a benign schwannoma is good and recurrence is rare after complete excision.

II. Case Presentation:

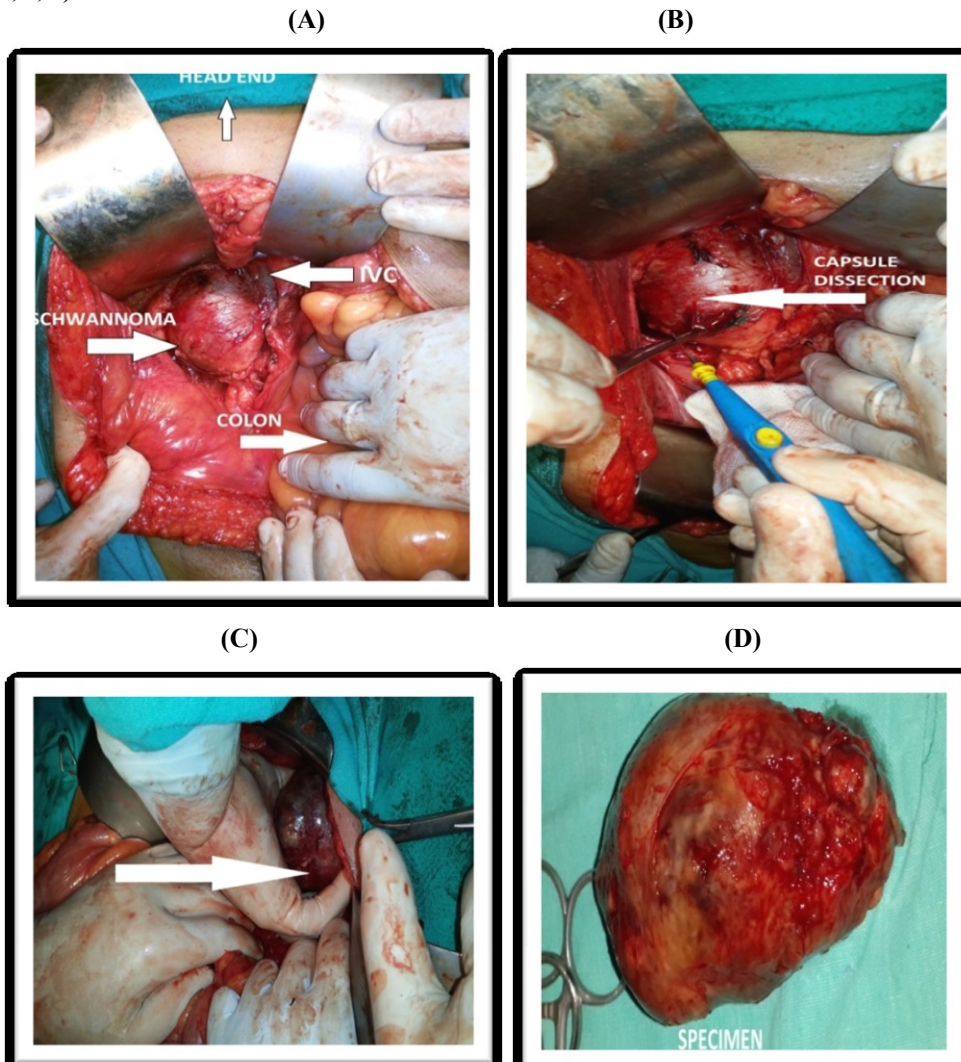
Authors report a case of 69 yrs old male patient resident of Ranbirsinghpura, Jammu retired army personnel who reported to our centre with chief complaint of mild intensity pain over central and right lower abdomen over a period of 3 months. Pain was insidious in onset, non radiating, continuous and dull aching in character. Bowel and bladder habits were normal. No history of haematemesis, melena, jaundice, prolonged fever and haematuria. Patient was a known diabetic and hypertensive for 6 years on medications. No similar pain complaints in the past nor any other family member had similar complaint. On clinical examination the patient was afebrile, pale and comfortable. Blood pressure and heart rate were normal. Abdominal examination was unremarkable with no abdominal mass palpable, no tenderness no hepatosplenomegaly with normal bowel sounds. Digital rectal examination was also normal. The chest X-ray, haemogram, biochemical analysis were all within the normal limits. Abdominal ultrasound and contrast enhanced CT scan revealed well defined heterogeneous contrast enhanced soft tissue density mass lesion measuring 12 x 7 x 5 cm at right paraspinal region with central necrosis [Fig 1A,B]. The mass was localized just above the right psoas muscle and inferior to the right kidney, slightly compressing the inferior vena cava. CT guided FNAC was planned but could not be done due to patient's disapproval. In view of above findings surgeons were confused about the type of neoplasm, about its malignant status and whether resectability would be possible due to its proximity to great vessels. A decision was reached that if the mass would be invading adjacent structures and intra-operatively features suggested malignancy, resection of wide local excision would not be attempted and only biopsy specimen would be taken for histopathology and next step for management would be decided after histology report. On the contrary on surgical exploration, a large encapsulated non invasive mass was found which was dissectible from the adjacent structures, it was very carefully dissected from inferior vena cava as it was almost impinging the large vein [FIG 2A,B,C,D]. It was possible to remove the complete tumour together with its capsular lining but no peripheral nerve entering or exiting the tumour was definable. Pathological and immunohistochemical

evaluation revealed it to be a benign schwannoma (ancient variant). Postoperative and follow up period was uneventful .

(FIG 1A, B) CECT ABDOMEN



(FIG 2A,B,C,D) INTRAOPERATIVE IMAGES



III. Discussion:

Schwannoma is a neurogenic tumor usually arising between the third and sixth decades of life, with a predilection for men. It may occur in any organ or nerve trunk with the exception of cranial nerves I and II which lack schwann cells. A schwannoma typically appears as a solitary, ovoid or spherical mass with well-defined borders, in large tumors (>8-10 cm) we may find degenerative pattern (cystic areas, calcifications, interstitial fibrosis, hyalinization) which is predominant in the "ancient" variant, a subtype of the typical or classic schwannoma with a very favorable clinical outcome. Early symptoms of retroperitoneal tumors are bizarre and none of the symptoms which occur can be considered diagnostic. Reported symptoms are vague, poorly localized abdominal pain and discomfort, accompanied by non-specific digestive disturbances. Referred pain and neurological symptoms in the lower extremities have also been described. The differential diagnosis for retroperitoneal schwannoma includes other neurogenic tumors such as paraganglioma, pheochromocytoma, liposarcoma and malignant fibrous histiocytoma. Hence the diagnosis of a benign retroperitoneal tumour is mainly one of exclusion, but ultimately based on histology. Therefore, misdiagnosis of retroperitoneal schwannomas is not uncommon. CT-guided core biopsy and fine needle aspiration have been found to be unreliable for the diagnosis of retroperitoneal schwannoma[4]. They may be helpful only if the sample contains enough schwann cells to visualize microscopically. However, in areas of degeneration, the cellular pleomorphism can hinder the diagnosis, and degenerative cells may be misinterpreted as malignancy. One also runs the risk of hemorrhage, infection, and tumor seeding; thus, many authors do not recommend CT-guided biopsy. Therefore, surgical resection is the only accurate approach for pathologic evaluation to enable diagnosis of retroperitoneal schwannoma. Wide surgical resection in cases of benign retroperitoneal schwannomas has been advocated by some authors based on their belief that malignancy can never be totally excluded. Others believe that because this is a benign mass, a simple enucleation or partial excision of the tumor is sufficient. The argument here is that the morbidity associated with resection of adjacent tissue would not be justified in the treatment of a benign lesion[5]. Chemotherapy and radiotherapy have only limited role with marginal added benefit in malignant schwannomas[6]. The common pathological variants of schwannoma are (a) Conventional schwannoma (b) Ancient schwannoma (c) Cellular schwannoma (d) Plexiform schwannoma (e) Melanotic schwannoma and other rare variants[7]. Typical schwannomas are composed of inter mixed Antoni A components (cellular and arranged in short bundles or interlacing fascicles) and Antoni B areas (less cellular and organized with more myxoid components). Characteristically all schwannomas show uniform and intense staining for S 100 protein.

IV. Conclusion:

The present case report emphasizes that surgeons should be aware of this rare neoplasm i.e retroperitoneal schwannoma and its various non specific clinical associations and ways of presentations. Authors expressed the diagnostic and management dilemma which they faced and how they proceeded, perhaps this would aid other surgeons to manage such neoplasms in future.

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Ethical Approval Statement:

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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