MASSIVE RETROPERITONEAL MASS – GIANT SCHAWANOMA

MUHAMMAD TAIMUR[1], SUNDAS ISHTIAQ[2], ABDUL BASIT[3], ABDUL HALEEM[4]

ABSTRACT

Schwannoma are usually solitary, slow growing, and non-aggressive neoplasms usually found in head and neck region. Retroperitoneal Schawanoma especially large sized are rare and difficult to diagnose clinically. We are discussing a 43 years old lady, who presented with huge reteroperitoneal mass of left side, treated successfully.

KEY WORDS:

INTRODUCTION

Schwannomas are benign nerve sheath tumours. The malignant schwannomas have been reported in literature but they frequently originate from the conversion of plexiform neurofibromatosis rather from the malignant degeneration of a benign schwannoma [1,2].

Principally, large retroperitoneal schwannomas are extremely rare tumours. They usually reach a large size when they arise retroperitonealy, which might raise suspicion for malignancy. Such tumors predominate in women [3,4]. Due to atypical or non-specific clinical presentation or lack of specific laboratory data, a preoperative diagnosis and tumour's origin is difficult to achieve or define respectively[3]. Nevertheless, the mainstay of management remains surgical removal and alternatively, the radio-therapy may prove to be effective[5]. Regarding prognosis[5], 10 and 20-year survival among patients having onset of symptoms at younger age (<25 years) is 80%, 60% and 28%, respectively. In contrary, the rate of survival among patients with onset of symptoms more than 25 years of age is 100%, 87% and 62%, respectively. Better survival rates are reported in patients with smaller size of tumor at the time of diagnosis (<2 cm in diameter)[6].

This case report is based on such a patient in whom the diagnosis was established with great difficulty and per-operative findings were something we had never encountered before.

CASE REPORT

A 43 years old lady presented with one year history of mass in left side of her abdomen. It was initially small in size and painless. At the time of presentation, the mass has increased in size and was crossing the midline. Patient had no symptoms regarding her urinary or gastrointestinal systems. Physical examination revealed a visible bulge in left hemi-abdomen and a palpable non-tender, firm to hard mass with well defined edges, smooth border and smooth surface, about 25 cm x 20 cm in dimensions. Upper limit of the mass was under left costal margin and was not palpable. Ultrasonographic examination revealed 25 cm x 20 cm heterogeneous mass arising from left half of abdomen and crossing the midline. Left kidney was not separately visible. Computerized tomography of abdomen showed retroperitoneal mass lesion on the left side also involving the left adrenal gland (Fig- 1,2). Trucut biopsy report showed metastatic poorly differentiated carcinoma. To get a more representative sample, incisional biopsy was performed which suggested spindle cell lesion with differential diagnoses of schwannoma with ancient change, fibromatosis and neurofibroma. Immunohistochemistry studies were positive for S-100 i.e. Neurofibroma. The patient was operated. Per-operatively, the mass was found to be adherent to descending colon, left kidney and ureter and left hemi diaphragm. The descending colon was separated. Left kidney and ureter were not salvageable as a result nephrectomy was performed. The mass was excised completely which weighed 12 kilograms (Fig-3). Diaphragm was repaired and a chest drain was placed. Post-operative recovery was uneventful.

1. Senior Registrar
2. House Officer
3. Postgraduate Trainee
4. Associate Professor
Department of Surgery
Fauji Foundation Hospital, Rawalpindi

Correspondence to:
Sundas Ishtiaq
House officer, Department of Surgery
Fauji Foundation Hospital, Jhelum Road, Rawalpindi
E-mail: sundas_ishtiaq@hotmail.com

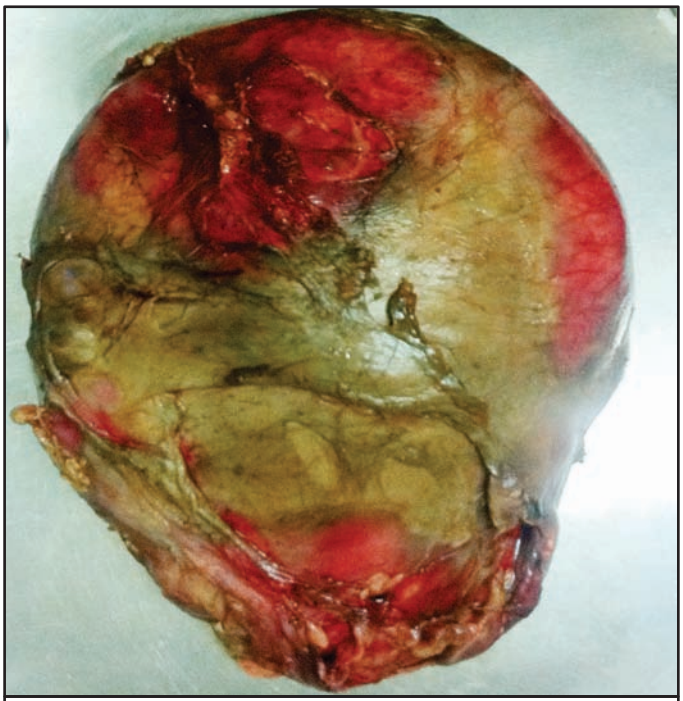

FIGURE-3: GROSS SPECIMEN OF TUMOUR AFTER SURGICAL EXCISION.

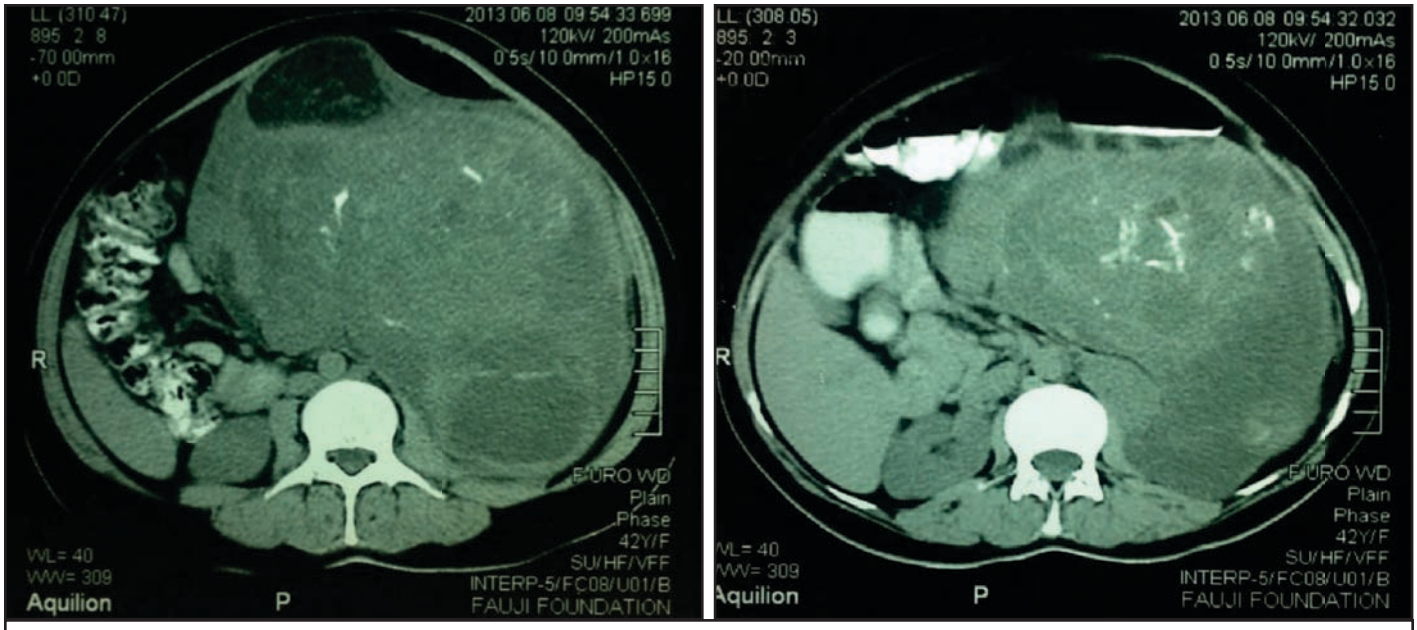

FIGURE-1&2: A LARGE HETEROGENEOUS RETEROPERITONEAL MASS CROSSING THE MID LINE, REACHING UP TO ANTERIOR ABDOMINAL WALL AND OCCUPYING THE WHOLE LEFT PARA-COLIC GUTTER AND PERITONEAL CAVITY.

DISCUSSION

Soft tissue sarcomas (STSs) account for less than 1% of all cancer diagnoses in the general population[7]. They occur more commonly in children, representing approximately 7% of all cancers in patients younger than 20 years of age[7,8]. Rhabdomyosarcomas comprise approximately half of the paediatric STSs while the remaining are a heterogeneous group of neoplasm known as non-rhabdomyosarcoma soft tissue sarcomas (NRSTSs)[8]. The Malignant Peripheral Nerve Sheath Tumours (MPNST) is defined as any malignant tumour differentiating into or deriving from the peripheral nerve sheath cells, with nonspecific symptoms, having high risk of recurrence locally and distant metastasis[5,8]. MPNST is particularly rare, affecting general population with an incidence of 0.001% and extremely rare spindle cell sarcoma in children, accounting for approximately 5-10% of non-rhabdomyosarcoma soft tissue sarcomas [8,9].

Schwannoma are tumours that are arise from the Schwann cells of peripheral nerve fibre and are generally found in the head, neck and extremities[10]. The capsule of these tumours is derived from epineurium[11]. Around 5 - 8% cases are associated with Von Recklinghausen's disease[6]. They are usually benign in nature and more frequently occur in adult females between 20-50 years of age (2:3, male to female ratio). Retroperitoneal schwannoma are more commonly present in the paravertebral spaces and pre sacral region[12]. If present in such locations, it is usually in association with Von Recklinghausen's disease. Among schwannoma that occur retroperitonealy, around 0.7% are benign and 1.7% were reported to be malignant yet producing vague symptoms such as abdominal pain and distension[11]. The Schwannoma are predominant in men among the age group 20 to 50 years. The commonest site is head and neck region and flexor tendon sheaths of extremities but sites may vary. The tumor at unusual locations such as the fallopian tubes, scrotum or urinary bladder has also reported in literature. Schwannoma in pelvis are very uncommon and comprises of only 1% of the cases[13]. These pelvic tumours usually present with non-specific symptoms like pain, rectal dysfunction or with palpable mass. Literature has also reported Schawanoma within the lumen of the gastrointestinal tract. These Peripheral Nerve Sheath Tumours comprise of 2- 6% of the Gastrointestinal Stromal Tumours (GISTs), which are commonly found in stomach and small intestine. Among colorectal schwannoma, the caecum is the most frequent site followed by sigmoid colon and recto sigmoid junction. They commonly present as abdominal pain, bleeding per rectum and colonic obstruction. Such colonic lesions may mimic as carcinoma and rarely the coexistent schwannoma with synchronous adeno-carcinoma in colon have been reported. Only few cases of rectal schwannoma have also been reported in literature [1,2].

MPNSTs exhibit a wide range of biological behaviour that ranges from low to high grade malignancy and a diversity of clinical manifestations; hence, the genetic and molecular findings in these tumours reflect a heterogeneous mixture of several aspects of MPNSTs[5,8]. A review of 120 MPNSTs at Mayo clinic during 71-year period demonstrated that patients with von Recklinghausen's disease have an incidence of 4.6% as compared to the incidence (i.e. 0.001%) among the general population without the disease. MPNSTs most often present as an enlarging palpable mass while complaints of pain are variable[8].

Grossly, the schwannomas are encapsulated, well circumscribed and frequently undergo cystic degeneration. Histologically, nuclear palisading may be a striking feature. Typical schwannoma comprise of inter mixed Antoni- A components (i.e. arranged in short bundles and cellular or with interlacing fascicles) and Antoni- B areas (i.e. less cellular, organized with more myxoid components). Contrary to it, the cellular variant of Schawanoma (i.e. large pelvic or retroperitoneal) has uniformly arranged spindle cell without

Antoni A or B areas. All schwannoma characteristically demonstrate intense and uniform staining for S-100 protein[5,14]. These are more aggressive and are likely to have a high incidence of local recurrence after excision. Chances of distant metastasis are also high. Malignancy cannot be ruled out pre-operatively or by frozen sections, hence, it is recommended that complete excision with negative margins is achieved. Recurrence usually occurs within six months of surgical excision and recurrence rates vary from 16% to 54%[12].

Schwannoma are usually solitary, slow growing, and non-aggressive neoplasm[6]. Majority of these tumours are asymptomatic and usually diagnosed incidentally or may present clinically with non-specific or vague symptoms[2]. On routine clinical examination, they are usually detected as an abdominal mass as in this case. The predominantly cystic nature of the tumour can demonstrate by ultrasound and CT scan examination but the exact nature cannot be diagnosed[1,9]. Malignant transformation is rare and malignant schwannoma are large in size and are highly aggressive. They are painful, and may cause many different symptoms depending upon the location and size of the tumour[13].

The investigation of choice in the diagnosis is MRI which gives a confirmation of the origin, extent, internal structure and infiltration of the lesion. The characteristic diagnostic features on MRI are "Target sign" which is hypo-intense centre with hyper-intense periphery, and "Fascicular sign" which is seen in presence of fascicular bundles. In retroperitoneal schwannoma, these findings are not present which make the diagnosis quite challenging. Ultrasonography is useful in taking guided biopsy while CT scan helps in detecting associated malignant changes such as bony erosions[3]. If malignant change is present, irregular margins with infiltration into adjacent structures are seen on the scans[10,12]. In a study carried out in Japan, 94 (72.3%) cases of schwannoma turned out to be benign and 36 (27.7%) were found to be malignant[12].

Quite infrequently schwannoma's undergo malignant transformation invading neighbouring organs for instance colon, kidneys, and adjacent viscera[1]. Tissue diagnosis is established with FNAC and excision biopsy [9]. CT guided biopsies and FNAC are not always reliable as it is required that the sample contains a significant amount of Schwann cells. FNAC from the areas of degeneration may lead to a misdiagnosis of malignancy. At the same time, FNAC increases the risk of haemorrhage, infection, tumour seeding and as a result, it is not the preferred choice of many surgeons. There is a lack of specific standardised criteria for diagnosing malignant changes except for certain characteristics such as dense fascicules in marble like pattern comprising of asymmetrically tapered spindle cells[7]. Microscopically dense cellular areas are termed as "Antoni A" areas and necrotic hypo dense areas as "Antoni B" areas. Presence of mitotic figures, pleomorphism, and infiltration of blood vessels are usually characterised as being malignant [1].

MPNSTs are commonly metastasize to the lungs, followed by bone and pleura respectively [6]. Keeping in view this, a chest CT scan (computed tomography) is the preferred imaging to screen for distant metastatic disease[7]. Bone scan is also helpful to exclude any metastatic bone disease [8].

Care must be taken at the time of attempting removal of retroperitoneal and intra-pelvic Schwannoma. A thorough preoperative planning and involvement of specialists of other sub-specialities should be considered since complications have to be foreseen. Meticulous haemostasis can be a problems, if the capsule is adherent to the pre-sacral venous plexus. Moreover, hypervascularity of the tumour as reported in the literature, can further complicate its excision, and may warrant preoperative performance of an arteriogram, in cases of any doubt [15]. Wide surgical resection has been advocated by a few authors in cases of benign retroperitoneal schwannoma, based on their belief that malignancy can never be totally excluded. Local tumour excision should be regarded as the treatment of choice, since tumour recurrence or malignant transformation almost never occurs in benign schwannoma. Even in challenging cases where complete removal of the tumour was unattainable and simple enucleation was performed, no tumour enlargement or malignant change was observed later on[13,15].

Due to their insensitivity to radio and chemotherapy the malignant schwannoma carries a poor prognosis. Immuno-histochemical positivity for S-100 protein is a constructive diagnostic finding. Combinations of clinical, pathological and immune-histochemical studies usually help establishing a final diagnosis [13,14].

The cornerstone of treatment of MPNST remains wide and complete surgical excision in order to achieve negative margins as this offers the best outcome with respect to both local recurrence and distant metastases. Both adjuvant chemotherapy and radiation are also often utilised despite its limited response[1,9]. Preoperatively and post operatively the chemotherapy can be administered. Several cycles of adjuvant chemotherapy with cyclophosphamide/doxorubicin have been advocated, according to the literature[8].

In cases when the postoperative histology gives evidence of the tumour being malignant after marginal excision, local recurrence has to be expected in up to 72% of cases. However, recurrence of the tumour after resection involving wide surgical excision margins has been reported in only 11.7% of the cases 3. In cases of unexpected event of proven malignancy, re-resection should be considered if a wide margin has not been achieved initially[15].

CONCLUSION

Schawanoma has a poor prognosis because of their insensitivity to chemotherapy and radiotherapy so early diagnosis and complete excision is the mainstay of treatment to improve prognosis.

REFERENCES

1. Q Li, C Gao, J T Juzi, X Hao. Analysis of 82 cases of retroperitoneal schwannoma. Australian & New Zealand J of Surg. 2007;77(4):237-40.
2. Samarakoon L, Weerasekera A, Sanjeewa R, Kollure S. Giant pre-sacral schwannoma presenting with constipation: a case report. J of Med Case Reports. 2012;6:285

3. Sakalauskaite M, Stanaitis J, Saulius C, Pleckaitis M, Lunevicius R. Retroperitoneal giant schwannoma eroding lumbar vertebra: a case report with literature review. Cent Eur J Med. 2008; 3:233-44.
4. Fass G, Hossey D, Nyst M. Benign retroperitoneal schwannoma presenting as colitis: a case report. World J Gastroenterology. 2007; 13:5521-24.
5. Evans G. Neurofibromatosis type 2 (Nf2): a clinical and molecular review. Orphanet J Rare Dis. 2009; 4:16.
6. Patrinou A, Malindretos P, Koutroubas G, Anagnostou N, Argiraki E, Syrganis C. A rare retroperitoneal schwannoma in a patient with neurofibromatosis Type 2. Clinical Kidney J. 2010; 3(3):288-90.
7. Cury J, Coelho RF, Srougi M. Retroperitoneal schwannoma: case series and literature review.Clinics.2007;62:359-62.
8. Alavi S, Arzanian MT, Nilipour Y. Retroperitoneal Malignant Peripheral Nerve Sheath Tumour Replacing an Absent Kidney in a Child. Case Reports in Oncological Medicine. 2013; 2013: 234-38.
9. Tortorelli AP, Rosa F, Papa V. Retroperitoneal schwannomas: diagnostic and therapeutic implications. Tumori 2007; 93:312-15.
10. Wong CS, Chu TY, Tam KF. Retroperitoneal Schwannoma: a common tumor in an uncommon site. Hong Kong Med J. 2010; 16:66-68.
11. Kalayci M, Akyuz U, Demirag A, Gurses B, Ozkan F, Gokce O. Retroperitoneal Schwannoma: a Rare Case. Case Reports in Gastrointest Med. 2011; 2011:462-65.
12. Siddha K, Vijayan SKL, Shetty S, Bhat SR, Shetty S, Khadilkar UN. Retroperitoneal Schwannoma: An Atypical Presentation. J of Clin & Diagnostic Research. 2014;8(10):22-23.
13. Tauro LF, George C, Kamath A, Lobo G, Shetty P, Hegde BR. Giant schwannoma in the pelvic retroperitoneum. J of Clin & Diagnostic Research. 2008; 2: 1210-14.
14. Hooja1 N, Rajoria1 L, Verma1 A, Jindal A, Himani S, Malviya K. Retroperitoneal Schwannoma- A Case Report. Sch J App Med Sci. 2013; 1(5):479-81.
15. Schindler OS, Dixon JH. Retroperitoneal giant schwannoma: Report on two cases and review of the literature. Journal of Orthopaedic Surgery 2002;10 (1): 77-84