Retroperitoneal giant schwannomas: Report on two cases and review of the literature

Oliver S Schindler and John H Dixon
Bone and Soft Tissue Tumour Department, Avon Orthopaedic Centre, Bristol, England

Patrick Case
Orthopaedic Research Laboratory, Avon Orthopaedic Centre, University of Bristol, England

ABSTRACT

The occurrence of massive retroperitoneal schwannomas is extremely rare and their presence may only be expressed by insidious onset of non-specific and misleading symptoms with a predominance of lower back pain. MRI scan as the imaging procedure of choice will demonstrate the tumour location and its relation to the surrounding structures, but due to heterogeneity and degeneration in some tumours, it may mimic malignancy. Hence tissue sampling through needle biopsies are essential to verify the diagnosis prior to surgery.

Tumour excision in toto is considered the treatment of choice, but it can be hazardous especially if the tumour is adherent to the presacral venous plexus. Severe bleeding complications due to the damage of venous structures have to be encountered, and establishing lasting haemostasis may pose considerable difficulties. Hence surgery should be attempted with full precautions, and preoperative counseling of the patient. If malignancy can safely be excluded, laparoscopic ‘piecemeal’ excision should be considered as an alternative treatment as recurrence is unlikely. Definition of the originating nerve might not always be possible and a minor degree of neurological impairment has therefore to be anticipated.

Key words: benign retroperitoneal tumour, giant, ancient, schwannoma, sacral plexus bleeding, surgical complications

INTRODUCTION

Benign schwannomas are generally slow growing and painless tumours originating from the Schwann cells of peripheral nerve sheaths and predominantly occur in females between the 2nd and 5th decade of life.8, 9, 27 As they are well demarcated by a thick capsular lining and not growing invasively, local excision is considered the treatment of choice.9 Once completely excised, recurrence of benign schwannomas is not expected. Adjuvant therapy therefore will not be administered.2, 26, 30

Schwannomas do not usually exceed a diameter of 5 to 6 cm,34 but larger size tumours up to a diameter of 28 cm have been reported.16, 21, 29 Although Schwannomas occur most often in superficial layers at peripheral nerve sites, cases of retroperitoneal...
tumour locations have been described.\(^1\)\(^{,}\)\(^{24}\)\(^{,}\)\(^{32}\) The diagnosis in those cases is often delayed due to the absence of clinical signs until the tumour enlarges to a size capable of causing compression of intra-abdominal or intrapelvic organs.\(^7\)\(^{,}\)\(^{17}\)\(^{,}\)\(^{25}\)

We are reporting two cases of large expanding retroperitoneal Schwannomas. The necessity for needle biopsy is emphasised, as none of the currently available imaging modalities provide sufficient information to exclude malignancy. Problems of surgical tumour removal are discussed and possible hazards highlighted.

**CASE ONE**

A 40-year-old woman presented with a six-month history of continuous lower back pain independent of activities without any signs of referred pain or neurovascular deficit. Around that time she also started to suffer from frequency and a minor degree of urinary incontinence but did not pay any attention to it. Radiographs and Tc\(_{99}\) bone scan of the lumbar spine and pelvis were unremarkable. Non-steroidal anti-inflammatory drugs (NSAID) and physiotherapy provided some short-lived amelioration before she developed more severe signs of urinary incontinence, constipation, and discomfort during sexual intercourse. Clinical examination revealed a painless swelling in the lower abdomen, which raised the suspicion of neoplastic growth.

CT and MRI scans confirmed the presence of a large, retroperitoneal tumour mass located inferior to the promontorium and overriding the anterior aspect of the sacrum but not involving the adjacent viscera (Fig. 3–6). The tumour was oval in shape measuring 35x25cm and was causing ureteral, bladder and bowel deviation while the uterus and ovaries were pushed into the pouch of Douglas (Fig. 4, 5). A very shallow shoulder of the tumour extended into the left sided S\(_{1–2}\) foramina. Although the tumour mass appeared well defined and encapsulated, central areas of heterogeneity were visible suggesting possible malignancy (Fig. 4–6). Histology of a CT guided needle biopsy was in keeping with a benign ‘Ancient Schwannoma’.

Due to its size, its ongoing growth and increasing clinical symptoms, it was decided to excise the tumour by using a transabdominal/transperitoneal approach. It was impossible to trace the originating nerve, hence the complete specimen was retrieved by blunt dissection but not without damaging the presacral venous plexus. Haemostasis proved extremely difficult not least because of the limited access and poor visibility. After several attempts to stop the bleeding a compression pack was applied and the patient haemodynamically stabilised. 48 hours later the pack was renewed and after a further 48 hours the bleeding finally ceased allowing for closure of the wound. The overall blood loss estimated 24 litres and the patient received a total of 66 units of blood, 38 units of fresh frozen plasma (FFP) and 6 units of platelets (PLT). Clinically a vaginal sinus developed post operatively but resolved spontaneously, whilst a mild left sided hyposthesia around the S\(_{1–2}\) dermatome prevailed. The patient is left with a minimal degree urge/stress incontinence, which is currently under investigation and might be attributable to her operation.

Histological examination of the main resection specimen confirmed the diagnosis of the biopsy of an ‘ancient schwannoma’ surrounded by a thin true capsule (Fig. 1). Degenerative areas within the tumour were seen including cystic degeneration, large collections of foamy histiocytes, ischaemic foci with pyknotic cells and collagen replacement. Verocay bodies (palisading nodules) and Antoni A (highly ordered cellular component, well organized spindle cells in palisading pattern) and B (less cellular, loose textured pleomorphic cells and predominantly myxoid component) areas were present focally. There was hypercellularity in some areas though with a negligible mitotic rate (Fig. 2).

**Figures 1** Case 1: Photograph of the transected specimen showing heterogeneity of the Schwannoma with large central areas of tumour necrosis. Calcified areas can bee seen at the 3 o’clock and 9 o’clock position
CASE TWO

A 37-year-old man presented with insidious onset of left sided lower back pain. Clinical examination and radiographs appeared normal. He responded well to NSAIDS and back muscle exercises, but attended his GP 18 months later presenting a flare up of his lower back symptoms together with a dull lateral abdominal pain radiating into his antero-lateral thigh. The pain increased in supine position and was superimposed by the occasional colic.

Abdominal ultrasound revealed a large encapsulated soft tissue mass of oval shape measuring 20x12cm in size, it was characterized by a solid shell with some through transmission and numerous well defined fluid areas of varying size within it. A CT scan with and without contrast (Gadolinium) was inconclusive raising the suspicion of a haematoma or a soft tissue sarcoma (Fig. 7, 8). Subsequent MRI scans confirmed the encapsulated status of the tumour, which was lying in the retroperitoneum displacing the ilio-psoas muscle distally and the left kidney cranially (Fig. 9–11). No hydronephrosis of the ipsilateral kidney was seen. A soft tissue needle biopsy revealed the presence of a benign Schwannoma.

The tumour was approached through an oblique skin incision in the left axillary line. It was possible to remove the complete tumour together with its capsular lining, but no peripheral nerve entering or exiting the tumour was definable. A total blood loss of 1200 ml was estimated but no substitution of blood products was necessary. No complications were encountered during surgery but the patient presented post-operatively with a transient partial femoral nerve palsy characterized by mild anterolateral thigh paraesthesia (L₃-dermatome) and MRC (Medical Research Council Rating Score) grade IV+ quadriceps muscle power. At six months his quadriceps weakness was completely restored and the paraesthesia, although still present, was markedly improved. Histology findings were similar to the ones described for case 1, but without the same degree of hypervascularity.

DISCUSSION

Historically schwannoma, perineural fibroblastoma and neurinoma were expressing the opposing views regarding the histogenesis of so called nerve sheath tumours. Structural evidence has decided in favour of schwannoma or neurilemmoma as the tumour is defined as a benign neoplasm arising from the myelinated nerve sheaths. It predominantly consists of Schwann cells characterized by their palisading architecture. The name neurinoma, introduced by Verocay, literally means nerve-fibre tumour and is therefore clearly inappropriate.

Only 0.3–3.2% of benign schwannomas are found in retroperitoneal locations. As those tumours are usually painless symptoms occur due to expansion and hence diagnosis and treatment are delayed. The presentation of unspecific lower back pain in middle aged patients are too often regarded as an epidemic disease affecting the great majority of the population by one or more episodes in life. Care must be taken by evaluating the patient’s history in view of signs and symptoms not directly related to the painful area. Although the occurrence of presacral schwannomas is extremely rare, an abdominal examination should always be incorporated in a thorough examination when investigating a patient for lower back pain or sensory disturbance around the buttocks and the saddle area. As in the presented cases this seems to be of particular importance where insidious onset of symptoms is present.

Foote and co-workers stated in their article in 1963 that early symptoms of retroperitoneal tumors are bizarre and none of the symptoms which occur can be considered diagnostic. Reported symptoms are vague, poorly localized pain and discomfort, accompanied by non-specific digestive disturbances. Referred pain and neurological symptoms in the lower extremities have also been described, and in one case the patient presented with varicose veins. Hence the diagnosis of a benign retroperitoneal tumour is mainly one of exclusion, but ultimately based on histology.
Figures 3 Case 1: Axial CT scan displaying a large abdominal mass of slightly mixed attenuation, lying within the pelvis and posterior to the rectum. The mass is in close relationship to the rectum but no obvious bony destruction is seen.

Figures 4 Case 1: Axial T1-WI confirming the retroperitoneal position of the tumour, which is well defined and encapsulated. It occupies the whole concavity of the sacrum without causing any sacral destruction. The iliac vessels particularly on the left are displaced anteriorly and laterally by the tumour mass.

Figures 5 Case 1: Axial T2-WI (identical scanning sequence as in Fig. 4). The mixed signal of the tumour as seen here demonstrates more clearly than either CT or T1-WI the inhomogenous nature of the tumour. The periphery of the tumour enhances on T2-WI, representing viable tumour tissue, whilst the central areas show low signal intensity similar to T1-WI, in keeping with areas of tumour necrosis. A very small shoulder of the tumour appears to extend towards the left S₁/₂ foramina with continuation into the S₁ nerve root.

Figures 6 Case 1: Sagittal T1-WI with contrast (Gadolinium) demonstrating slight increase in signal contrast between central and peripheral areas of the tumour mass when compared to T1-WI. The sagittal view emphasizes the direct proximity of the tumour to the sacrum and its position within the pelvis.
Although the cases presented proved histologically to be truly benign Schwannomas, heterogeneity and cystic changes of schwannomas as displayed on MRI or CT scan have been reported as signs of malignancy especially in patients with underlying von Recklinghausen’s disease.\(^{19,36}\) Takatera and co-workers noticed the presence of cystic changes in 75% of malignant Schwannomas compared to only 6% in benign lesions.\(^{36}\) Heterogeneity is usually caused by cystic degeneration, hence the description as ‘ancient schwannoma’ (Fig. 1, 4–6). The degeneration is due to central tumour necrosis as the schwannoma grows to a size beyond the capacity of its blood supply in maintaining survival of central areas.\(^7\) Tumours presenting with these features should raise a high degree of suspicion by the treating surgeon and hence to be treated as potentially malignant unless proven otherwise. Calcifications within the tumour mass as seen on plain radiographs of benign retroperitoneal Schwannomas has only been described once in the literature and should not therefore be considered diagnostic evidence.\(^2\)

Computed tomography (CT) and magnetic resonance imaging (MRI) are widely used as imaging techniques in the evaluation of retroperitoneal soft tissue tumours. The diagnostic value of CT does however appear mitigated by its limited resolution and soft tissue definition, as demonstrated in one of the presented cases, where CT scans led to the suggestion of a haematoma (Fig. 7). CT images fail to adequately reproduce stroma heterogeneities, a main characteristic for ancient Schwanomas, when compared to MRI (Fig. 3, 7). However, addition of intra-venous Gadolinium as a contrast medium may provide enhancement of tissue inhomogeneities within the tumour (Fig. 8). Today, MRI is generally accepted as the imaging modality of choice for most soft tissue lesions. It provides higher diagnostic predictability than CT or ultrasound when imaging soft tissue tumours, and in return it may lead to a more accurate preliminary diagnosis (Fig. 4–6, 9–11).\(^4,19\) However it is incapable of reliably distinguishing between benign and malignant tumours. Schwannomas characteristically possess low signal intensity on T1-WI similar to muscle (Fig. 4, 9) and high signal on T2-WI similar to fat (Fig. 5, 6, 10). Beside standard T1 and T2 weighted images (T1-WI, T2-WI) we therefore recommend the

**Figures 7** Case 2: Axial CT scan displaying a large abdominal mass extending antero-laterally to the iliac wing and the anterior abdominal wall. It is well encapsulated and separated from the iliacus muscle by a distinct fat plane. The abdominal mass has got lower attenuation than muscle and shows slight patchy enhancement, which could possibly suggest some stroma within it.

**Figures 8** Case 2: Axial CT scan with Gadolinium (Identical scanning sequence as in Fig. 7). Patchy enhancement of the tumour, which could suggest the presence of stroma within it, is displayed following the application of Gadolinium.

**Figures 9** Case 2: Axial T1-WI showing a large retroperitoneal tumour of low signal intensity, displacing all adjacent structures. It is of slightly mixed attenuation without demonstrating any cystic lesions.
acquisition of a fat suppression sequence (STIR) on which the schwannoma will maintain its high signal allowing for delineation from pure lipomatous tumours (Fig. 11).

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. Since tumour recurrence or malignant transformation almost never occurs in benign schwannomas, local tumour excision should be regarded as the treatment of choice. Even in difficult cases where complete removal of the tumour was impossible and simple enucleation was performed no tumour enlargement or malignant change was observed. However malignant transformation (neurofibrosarcoma), although extremely rare, is usually only observed in cases with underlying von Recklinghausen’s disease. It is therefore of paramount importance to survey the patient for stigmata of von Recklinghausen’s disease at first presentation.

Should the postoperative histology confirm malignancy of the tumour local recurrence after marginal excision has to be expected in up to 72% of cases, whereas recurrence after resection with a wide surgical margin has been reported in only 11.7%. In such an unexpected event of proven malignancy one should probably consider re-resection if a wide margin has not been achieved originally.

Care must be taken in attempting removal of retroperitoneal and intrapelvic Schwannomas. Thorough preoperative planning and involvement of specialists of other subspecialities should be contemplated, as complications have to be anticipated. Haemostasis can pose problems if the tumour capsule is adherent to the presacral venous plexus. Moreover, hypervascularity of the tumour as reported in the literature can further complicate its excision, and may warrant in cases of doubt the preoperative performance of an arteriogram. Sufficient amounts of blood products have to be readily available including fresh frozen plasma and thrombocytes. The anaesthetist should be made aware that a high volume blood loss might be encountered.

In a case report by Foote the attempt to excise a large retroperitoneal schwannoma was abandoned because of the danger of uncontrollable hemorrhage. Carpenter reported one intraoperative death related to uncontrollable hemorrhage from severing the right common iliac artery during a difficult dissection. In his series 5 out of 21 cases with retroperitoneal ganglioneuromas were said to be inoperable and 3 patients consequently died due to complications related to their tumours. It has to be borne in mind that in the majority of cases problems with tumour resection are due to the vicinity of the tumour to the surrounding neuro-vascular systems rather than to its own vascular supply.

Definitive diagnosis is based on histological analysis of biopsy specimens. Immunohistochemical staining can further support the diagnosis, as benign Schwannomas stain characteristically with immunoperoxidase techniques for S–100 protein, which represents a neural protein within the Schwann cell. This can also help distinguish between Schwannomas and neurofibromas, since the latter react poorly to S–100 protein staining, due to their perineural origin.
CONCLUSION

Unspecific lower back pain might be the only presenting feature in retroperitoneal Schwannomas, causing delays in diagnosis and treatment. MRI is the imaging modality of choice in demonstrating tissue heterogeneity and anatomic location of the tumour. However, needle biopsy should be regarded as the diagnostic gold standard, as none of the currently available imaging modalities provide sufficient information to exclude malignancy. Surgical removal although considered the treatment of choice, can pose extremely hazardous in retroperitoneal locations. Hence, piecemeal excision should be given consideration, provided enough evidence about the benign nature of the tumour is available. Definition of the originating nerve may not always be possible and a minor degree of neurological impairment is therefore to be anticipated.

ACKNOWLEDGEMENT

The authors would like to express their appreciation for the support given by Mr. Ian Eyre-Brook, Consultant Surgeon and Dr. Adam, Consultant Pathologist of the Musgrove Park Hospital in Taunton/Somerset, by the Medical Photography Department at Southmead and Frenchay Hospitals in Bristol, and by Sylvia-Louise Davies.

REFERENCES