



A Giant Gluteal Schwannoma with extension into the Pelvis: A case report

Giant Gluteal Schwannoma avec l'extension dans le Bassin : un rapport de cas

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ABSTRACT

BACKGROUND: Schwannomas are uncommon slow growing tumours arising from the nerve sheath or Schwann cell.

OBJECTIVE: To report a case and the surgical removal of a giant complex schwannoma and to highlight the value of extensive investigations including a preoperative histologic diagnosis in the successful surgical management of uncommon large benign tumours.

METHODS: A 39-year old man presented with a 15-year history of a large mass in the right gluteal region. He was clinically evaluated, subjected to imaging studies and surgery.

RESULTS: Clinical examination revealed a 40cm by 60cm mass in the right gluteal region and continuous with a 25 cm by 15 cm pelvic mass. The ultrasound scan revealed a very complex (mixed solid and fluid containing) gluteal mass with extension to the pelvis. The CT scan showed a very large, well defined lobulated tumour with cystic spaces and enhancing nodules. The tumour extended through the ischiadic foramen into the pelvis and posteriorly into the thigh muscles. A Doppler scan of the pelvic vessels revealed that the right common and internal iliac arteries were both compressed but not occluded. An incisional biopsy was reported as a Schwannoma with xanthomatous changes and an immuno histochemistry profile of S-100+, Ki-67+ (less than 1% of the cells). A right foot drop following the surgery responded to physiotherapy. The duration of total hospital stay was 12 weeks.

CONCLUSION: The staged excision of large and complex schwannomas is safe. It is essential that a preoperative histological diagnosis is made to establish that the tumor is truly benign. WAJM 2009; 28(3): 185–188.

Keywords: Tumour; Schwan cells; Benign Tumour; Scnwannomas.

RÉSUMÉ

CONTEXTE: Schwannomas sont des tumeurs de croissance lentes rares émanant du fourreau de nerf ou de la cellule Schwann.

OBJECTIF: signaler un cas et l'enlèvement chirurgical d'un complexe géant schwannoma et accentuer la valeur d'enquêtes étendues en incluant un diagnostic histologic préen vigueur dans la direction (gestion) chirurgicale réussie de grandes tumeurs bénignes rares.

MÉTHODES: un homme de 39 ans présenté avec une histoire de 15 années d'une grande masse dans le droit gluteal la région. Il a été cliniquement évalué, fait subir au fait de refléter des études et une chirurgie (opération).

RÉSULTATS: l'examen clinique a révélé 40 centimètres par la masse de 60 centimètres dans le droit gluteal la région et continu avec 25 centimètres par la masse pelvienne de 15 centimètres. Le scanner d'ultrasons a révélé un très complexe (s'est mélangé ferme et le liquide contenant) gluteal la masse avec l'extension au bassin. Le scanner de CT a montré une tumeur lobulée très grande, bien définie avec les espaces cystiques et les nodules améliorants. La tumeur s'est étendue par l'ischadic foramen dans le bassin et postérieurement dans les muscles de cuisse. Un scanner de Doppler des vaisseaux pelviens a révélé que les artères iliac communes et intérieures droites ont été les deux comprimées, mais n'occlues. Une biopsie incisionale a été annoncée comme un Schwannoma avec les changements de xanthomatous et un immuno histochemistry le profil de S-100 +, Ki-67 + (moins de 1 % des cellules). Une goutte (baisse) de bon pied suite à la chirurgie (l'opération) a répondu à la physiothérapie. La durée de séjour d'hôpital total était 12 semaines.

CONCLUSION: l'excision organisée de schwannomas grand et complexe est sûre. Il est essentiel que l'on fait qu'un diagnostic histological préen vigueur établisse que la tumeur est vraiment bienveillante. WAJM 2009; 28(3): 185–188.

Mots clé: la Tumeur, les cellules de Schwan, Bienveillantes, Scnwannomas.

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Abbreviations: CT, Computed tomography; MRI, Magnetic resonance imaging.

INTRODUCTION

Benign schwannomas are slow growing painless tumours that arise from the Schwann cells of peripheral nerve sheaths. They occur mostly in females between the second and fifth decades of life.¹⁻² The tumours do not usually exceed a diameter of 6cm although larger sizes of up to 28cm have been reported.¹⁻³ Schwannomas occur more often in superficial layers of the body at peripheral nerve sites although they may be found at a more concealed site such as the retroperitoneum and the mediastinum.²⁻⁴ In these locations the diagnosis of schwannomas is often delayed as there are few clinical signs until the tumour assumes a largelarger size which it compresses ion the of intra abdominal and intra pelvic structures.¹⁻⁴ These tumours are truly benign as they are well demarcated by a thick capsule and do not invade surrounding structures.⁵ Complete local excision is the treatment of choice, and once excised completely local recurrence is not expected.⁶ Further adjuvant treatment is usually not required.

We present a report on the diagnosis and surgical management of a giant complex schwannoma in a two-staged operation.

Case Report

A thirty-nine-year old man presented with fifteen years history of a painless mass in the right gluteal region. He had presented himself because he was unable to sit and walk. On examination there was a 40cm by 60cm mass occupying and distorting the entire right gluteal region (Figures 1). The mass was not attached to the overlying skin but the posterior aspect was not well defined. Abdominal palpation revealed a fixed mass 25cm by 20 cm below the umbilicus. (Figure 1 white arrow). One could get above the mass but not below it. A clinical diagnosis of a soft tissue sarcoma involving the right gluteal region and the pelvis was considered.

Imaging Findings

A CT scan revealed a very large, well defined lobulated tumour with cystic spaces and enhancing nodules. The tumour appeared hyper-vascular and to



Figure 1: Giant right gluteal tumour will pelvic extension: (arrow).

be lying between the gluteal muscles, extending through the ischiadic foramen into the pelvis. The pelvic component was approximately 15cm in diameter and showed a left pelvic "satellite" of 5cm. The mass also extended far into the thigh posteriorly between all muscle groups. The deep femoral artery and vein were displaced anteriorly while the iliac arteries were displaced antero-laterally and compressed, but were not occluded. A bilateral lower limb vascular Doppler scan performed at the same time concluded that there was compression of the right common iliac vein by a supra pubic mass. The radiological diagnosis was a mass of neurogenic origin, most likely the sciatic nerve and suggested Schwannoma or neurofibroma. Histology from an incisional biopsy was reported as a Schwannoma with predominant granular cell differentiation and xanthomatous changes (Figure 2). The immuno

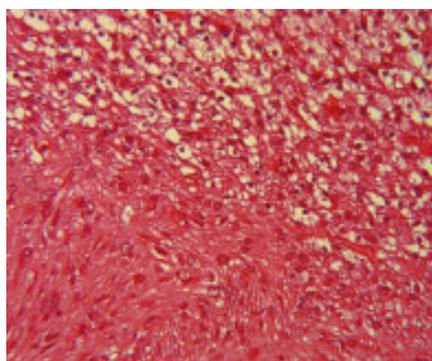


Figure 2: Schwannoma (Biphasic) Histology: The left lower section of the slide shows Antoni Type A areas of many highly ordered cells. The right upper section of the slide shows the Antoni Type B areas of fewer more loosely arranged cells and granular cell differentiation.

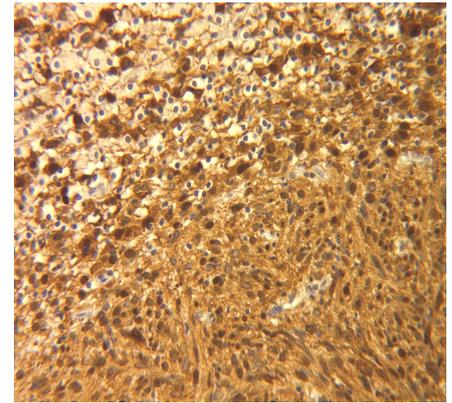


Figure 3: Schwannomas characteristically stain with immunoperoxidase techniques for S- 100 protein¹² which represents a neural protein within the Schwann cell.^{3,12}

histochemistry profile (Figure 3) was reported as S-100+. The proliferation marker Ki-67+ was reported as less than 1% of the cells confirming the benign nature of the schwannoma. The final diagnosis: was a benign Schwannoma most likely arising from the sciatic nerve.

Surgical Management

On account of the very large size of the mass and the pelvic extension, it was decided to excise the mass in two stages. The gluteal component was exposed through a -25 cm curved skin incision. It was impossible to trace the originating nerve; hence the complete specimen (Figure 4) was achieved by a combination of sharp and blunt dissection with consequent brisk haemorrhage from the gluteal muscles. Haemostasis was achieved with considerable difficulty by a combination of suturing, suture ligatures and diathermy. The patient was transfused eight units of whole blood. To prevent the possibility of a clostridia wound infection due to closeness to the perineum, ten mega units of penicillin G was administered daily intravenously for the first five postoperative days. In the immediate post operative period the patient developed a right foot drop. This improved on physiotherapy for twelve weeks. A second abdominal CT scan performed ten weeks after the first operation revealed a large multi lobulated non enhancing mass lesion in the right supra pubic region with displacement of the urinary bladder and the rectum to the left. The bowel loops were also pushed to the flanks. There was extension



Figure 4: The complete surgical specimen of the gluteal component of the complex schwannoma.

inferiorly and posteriorly through the pelvis into the sacral region suggesting a retroperitoneal mass.

The second stage of the operation exposed the pelvic component of the tumour via a lower laparotomy incision after a bladder catheter had been inserted. There was a urologist in attendance. The complete pelvic portion of the specimen was retrieved by a combination of sharp and blunt retroperitoneal dissection. As the tumour extended to the pre-sacral plexus of veins brisk haemorrhage was encountered. The bleeding was successfully controlled by a 2-0 silk ligature on the right internal iliac artery, warm packs and diathermy coagulation.

DISCUSSION

Superficial benign schwannomas have been reported more often than the deeply located ones.^{4,7} Schwannomas in the gluteal location are, however, uncommon. Determining the most appropriate treatment for such superficial benign tumours of the nerves is often a clinical dilemma.⁷ Resection of the tumour may lead to loss of nerve function and the formation of a painful neuroma. In this report the patient developed a foot drop after resection of the gluteal tumour.

It is more than four decades now since Foote and others reported from Brooklyn New York the staged excision of a giant retroperitoneal neurilemmoma (benign schwannoma) in a male patient.⁸ Over the years there have been a number of case reports of large or giant retroperitoneal schwannomas.^{2,9-12} In most of these reports the diagnosis of retroperitoneal schwannomas was delayed

primarily because it is rare to have schwannomas in the retroperitoneal region.^{2,8} Foote and his colleagues highlighted the non-specific nature of the early symptoms of retroperitoneal schwannomas and stressed that these symptoms are non-diagnostic.⁸ Schindler and other workers recently reported two cases of giant retroperitoneal schwannomas, they stressed the importance of backache in middle aged patients with otherwise normal clinical and radiological findings.² In the case of under discussion, the clinical presentation was for the gluteal (superficial) tumour mass with no symptoms related to the tumour in the pelvic retroperitoneum which was revealed at abdominal palpation.

Computed tomography (CT) and magnetic resonance imaging (MRI) are widely used as imaging techniques when evaluating large soft tissue tumours.⁹⁻¹¹ The diagnostic value of CT is reduced by its limited resolution and poor soft tissue definition often failing to reveal sufficiently the stromal heterogeneity typical of schwannomas.^{1-2,9-11} In one reported case CT findings led to suggestion that a schwannoma could be a large haematoma.² Improved resolution is often achieved by the use of intravenous contrast such as iopamino as was in the case of in the patient under discussion. MRI is the imaging of choice provided the service is available.⁹⁻¹¹ In many African countries, MRI is available only in the Teaching Hospitals limiting the management of these cases to the large centres only.

Definitive diagnosis of schwannomas is based on the histological analysis of biopsy specimens.¹⁻⁶ In our case an incisional biopsy provided enough material for a definitive diagnosis complete with immunohistochemical studies. The typical biphasic pattern microscopic features of Schwannomas (Figure 2) coupled with the characteristic areas of degeneration produces the stromal heterogeneity typical of CT and MRI images of Schwannomas.⁹⁻¹¹

Schwannomas characteristically stain with immunoperoxidase techniques for S-100 protein,¹² (Figure 5) which represents a neural protein within the Schwann cell.^{3,12} This property provides support for the diagnosis and can help

distinguish between Schwannomas and neurofibroma as the latter reacts poorly to S-100 protein staining.¹²

Wide local excision of retroperitoneal schwannomas has been advocated by some surgeons and pathologists who manage these tumours on the basis that malignancy can never be totally excluded.^{6,13} However tumour recurrence or malignant transformation almost never occurs in benign schwannomas.¹⁴⁻¹⁵ Provided the diagnosis of a benign schwannoma is established pre-operatively, local excision or in difficult cases simple enucleation is adequate as no tumour enlargement has been observed.^{2,12}

For the successful surgical removal of retroperitoneal or pelvic schwannomas careful and detailed preoperative planning is required. The patient must be carefully counselled regarding the risks of residual functional disability due to either nerve damage or loss of muscle mass. Many problems with tumour resection are due to the closeness of the tumour on to surrounding neuro-vascular bundles as well as the vascular supply of the tumour. The surgical team must include urologists and vascular surgeons if possible. The anaesthetist must have some experience in dealing with operations associated with high volume blood loss. A ligature on the internal iliac artery on the side of the tumour early in the dissection may limit excessive blood loss and prevent uncontrolled haemorrhage which may necessitate the premature termination of the operation.

Surgical removal is the treatment of choice for of large complex schwannomas. Staged excision – provided a pre-operative diagnosis to establish that the tumour is benign has been made, which – may reduce the high risk of uncontrolled haemorrhage and damage to surrounding structures.

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