GIANT PRESACRAL SCHWANNOMA

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Abstract

Presacral schwannomas are rare and account for only about 1% of cases. A 32 year old female presented with a large presacral schwannoma which extends along the greater sciatic notch, bilaterally. It was a large multiloculated cystic mass and turned out to be a degenerating schwannoma on histology. Demonstration of cystic nature on imaging helps in the early diagnosis of schwannoma, as most other retroperitoneal tumors do not frequently form cysts.

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Introduction

Schwannomas are the most common peripheral nerve sheath tumours. They occur mainly in head and neck region or in the extremities. Schwannomas occurring in the pelvis are rare[1] and account for only about 1% of cases[2]. They usually do not elicit any significant neurological deficits[3]. When present in pelvis, thorax or in retroperitoneum, they present mainly with pressure symptoms. We present a case of a complex pre sacral schwannoma, which extended along both greater sciatic notches. The mass was detected incidentally during a routine antenatal ultrasound scan.

Case Report

A 32-year-old female patient was found to have a pelvic mass during routine antenatal sonography during her first pregnancy. No interventions had been done at that time. She had an uncomplicated full term cesarean section. Ante-partum, peri-partum and post-partum periods were uneventful. She had no symptoms related to the mass at that time, however six months later she developed difficulty in passing urine and episodic constipation. Subsequently she started experiencing a pain radiating along the back of her thighs, especially on standing. She also developed dysmenorrhoea. There was no lower limb oedema or varicose veins. She did not have a family history of neurofibromatosis. She underwent computed tomography (CT) and Magnetic resonance imaging (MRI) of the pelvis for further characterization of the lesion.

Non contrast CT scan revealed a low density mass measuring 9cm x 7cm x 3cm in the pre-sacral region of the pelvis (Figure 1) without calcifications or haemorrhages in it. The mass showed enhancing internal septations in

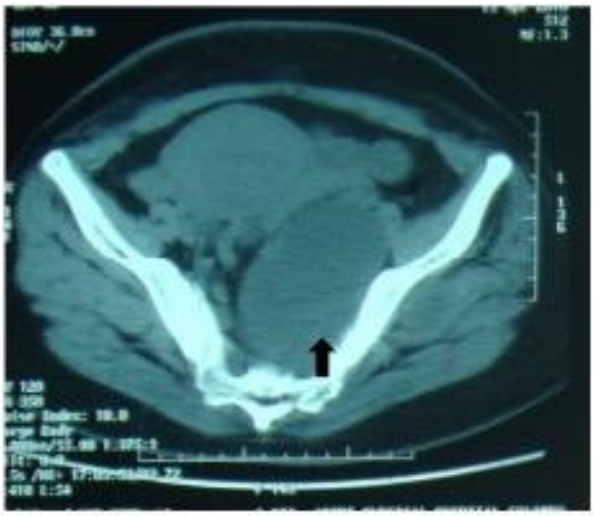

Figure 1. Non-contrast CT Pelvis Homogeneously low density mass lesion in the pelvis (black arrow)

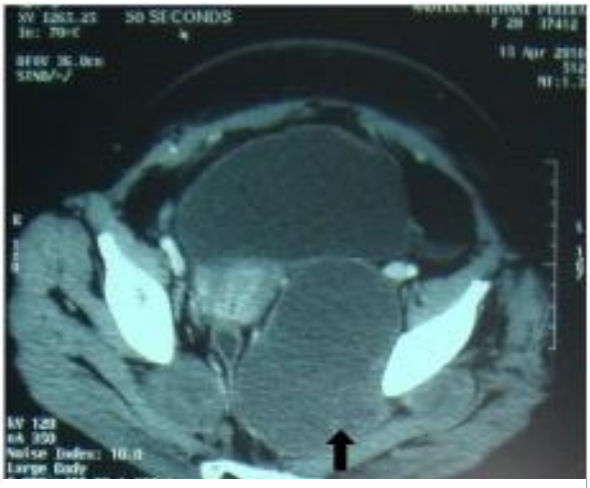

Figure 2. CECT Pelvis Mass lesion in the pelvis showing enhancing septa (Black arrow)

post contrast images (Figure 2). It was occupying almost the entire pelvis displacing the rectum right laterally, the uterus superiorly and the bladder anteriorly. There were two more lesions with similar characteristics in relation to both greater sciatic notches. However, communication with the main pelvic mass was not evident in the CT scan.

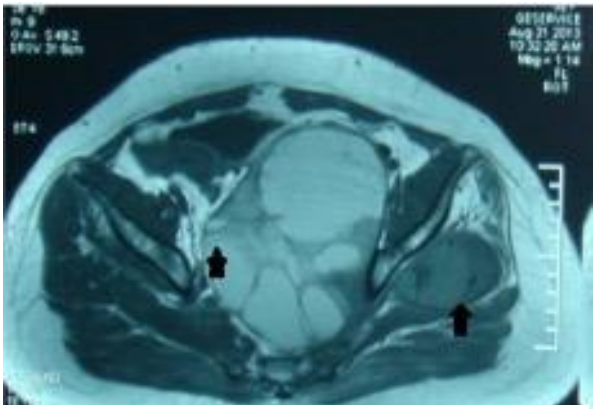

Figure 3. T1w MRI pelvis Large septated pelvic mass (black *) with another mass (black arrow) in relation to the left greater sciatic notch

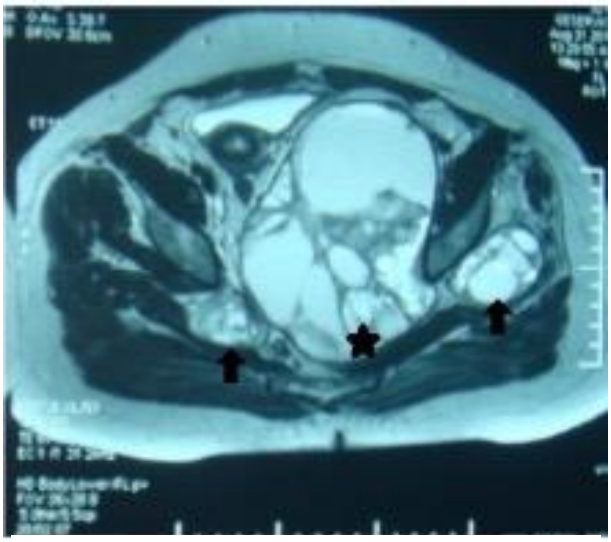

Figure 4. T2W MRI Pelvis Large septated pelvic mass (black *) with two masses (black arrows) in relation to each greater sciatic notch

MRI revealed a well-defined multiloculated cystic mass lesion in the pelvis with preserved fat planes between the mass and the pelvic viscera (Figure 3). There was no widening of the neural foramina or tumour extension to the sacral spinal canal. No communication of the presacral mass with the other two lesions was appreciated on MRI (Figure 4). She underwent surgery with a midline lower abdominal incision opening in to the peritoneal cavity and a large retroperitoneal cystic mass was identified. It was released from the surrounding structures without any damage to the surrounding viscera and was removed as a single mass along with the two other masses which communicated with the large mass. She had an uneventful recovery with no neurological deficits. Macroscopically the specimen showed an encapsulated mass measuring 9cm x 7cm x 4cm and cut sections showed cystic areas with papillary projections ranging from 1cm to 9cm in length.

Microscopy revealed that the mass was composed of spindle cells arranged in palisading bundles with absent mitotic activity. The histological diagnosis was degenerative schwannoma.

Discussion

Schwannomas are the most common type of peripheral nerve neoplasms, but are rarely seen in the presacral region[1]. Schwannomas can be either benign or malignant. The malignant schwannomas are usually associated with von Recklinghausen's disease[3]. MR imaging differentiates malignant schwannomas from benign ones by identifying indistinct margins and lack of a pseudocapsule. Giant pre-sacral schwannomas are thought to be originating from a peripheral nerve or adjacent to an anterior sacral foramen. Therefore, they are able to grow outside the confines of the bone with limited bone involvement. This results in minimal bone destruction or compression of the neurovascular structures. Therefore, the tumors are able to grow to a large size before presenting either as an incidental finding or with only minor symptoms[4]. This was observed in our patient as well where even after about one and half years of the

initial diagnosis she had minimal pressure symptoms and there was no bone involvement. Degenerative schwannoma, also known as ancient schwannoma, is characterized by cyst formation, hemorrhage, calcification, and fibrosis[3]. Preoperative diagnosis of schwannoma is difficult. But demonstration of its cystic nature in imaging helps in the early possible diagnosis as most of other retroperitoneal tumours do not frequently form cysts[5]. Cystic nature of this tumour was clearly depicted on MRI in our case.

This case highlights the importance of knowing the rare, but possible causes of presacral space masses such as schwannoma and the extensions of such lesions.

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