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CASE REPORT

Presacral ganglioneuroma: A case report and review of literature

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Abstract

Presacral ganglioneuromas are so rare benign tumors that only 17 cases have been reported in the literature. They are abdominal masses growing slowly and differential diagnoses have to be considered. Surgical resection is important for definitive diagnosis because it represents the only therapeutic choice. Because of the benign nature of ganglioneuroma, adjuvant chemo- or radiotherapy is not indicated but regular follow-up is necessary for an early diagnosis of potential local recurrence. We report a case of a 64-year-old man with a presacral ganglioneuroma.

Key words: Ganglioneuroma; Neuroblastoma; Presacral tumor

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INTRODUCTION

Ganglioneuromas are rare tumors in the neuroblastoma group[1-6]. They are benign lesions arising from sympathetic ganglion cells and complete surgical resection is considered to be curative[7-17]. They are rarely located in the presacral space, and so far, only 17 cases have been reported in the literature.

CASE REPORT

A 64-year-old healthy man sought medical advice in September 2005 because a routine abdominal ultrasonography for benign prostatic hypertrophy incidentally revealed a pelvic mass. He did not complain of any back pain or discomfort, and no neurologic symptoms were present. There was no evidence for endocrinological symptoms like diarrhea. He was admitted to our institute. Hereditary background for syndromes such as MEN2, cowden syndrome, tuberous sclerosis or familial polyposis coli were definitely excluded. Physical examination confirmed the presence of an abdominal mass, and neurologic examination was negative. There was no proctorrhagia, and both digital rectal examination and colonscopy excluded any origin of the pelvic mass from the rectum. Routine blood tests were made and tumor markers were detected (CEA, CA 19-9, CA 125, AFP, CA 72-4), but all values were normal. Laboratory studies showed no evidence of catecholamine excess. Computed tomography (CT) and magnetic resonance (MR) imaging were performed (Figure 1). Abdominal CT scan revealed a disomogeneous mass (12 cm × 9 cm × 8 cm) with irregular enhancement in the different contrastographic phases, arising from the sacral canal through the third right sacral foramen. Differential diagnosis had to be considered but a first diagnosis of Schwannoma or Chordoma was suggested by the radiologist. Pelvic MR confirmed the origin of the lesion from sacral canal in S2 through the third right sacral foramen, and excluded any sign of sacral or coccygeal metameric infiltration or osteolysis. Pelvic MR also confirmed the first hypothesis of Schwannoma. The patient was submitted to surgical laparotomy: midline incision and transperitoneal exposure were performed; small bowel, distal sigmoid and rectum and their mesenteries were retracted to expose the tumor lesion. Pelvis was completely occupied by a big mass, measuring 12 cm × 9 cm, tenaciously sticked to sacral plane. Intraoperative frozen section excluded the malignancy of the lesion. A partial resection of mesocolon and rectal preparation were necessary in order to gain access to the tumor lesion. Tumor preparation and its resection were very difficult because of large size, anatomical deep location and origin from the third sacral foramen. A complete and curative resection was performed. Histopathologic examination noticed ganglioneuroma with microcalcifications and focal lymphoplasmocellular infiltration. Large tumor ganglion cells are well differentiated and embedded in a neuromatous stroma with microcalcifications (Figures 2 and 3).

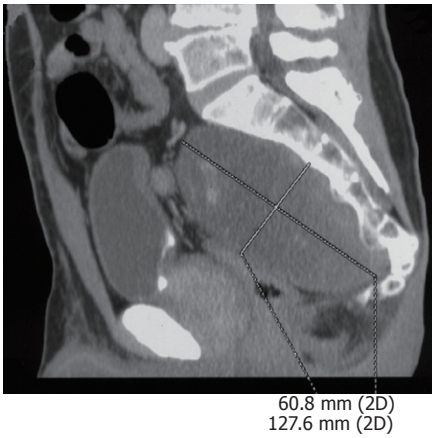

Figure 1 Abdominal CT scan shows a disomogeneous mass arising from the sacral canal through the third right sacral foramen.

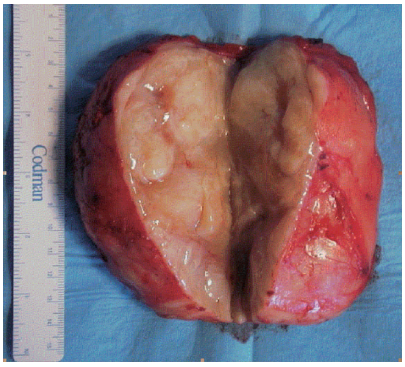

Figure 2 Tumor mass exposed after longitudinal section.

The patient had a regular postoperative hospital stay without complications and was discharged on the 7th postoperative day. After a 8-mo follow-up, he remains healthy with the exception of urinary retention, probably due to the benign prostatic hypertrophy.

DISCUSSION

Histologically ganglioneuromas are considered to be part of the neuroblastoma group together with neuroblastomas and ganglioneuroblastomas[1-6]. Ganglioneuromas are well-differentiated tumors, distinguished from the other groups because they are considered benign and constituted by mature sympathetic ganglion cells[1]. Some authors reported malignant transformation, spontaneously or after radiotherapy[15,18,19]. Arising along the sympathetic chain, ganglioneuromas are commonly localized in the posterior mediastinum followed by retroperitoneum, cervical region and adrenal gland[1,20]. Few cases have been reported in bone. The presacral location is very rare[7-14,17,21] (Table 1). An association with neurofibromatosis and multiple endocrine neoplasia syndrome Type Ⅱ B has been reported by other authors[6]. They are common in young females[6,22,23] and such as abdominal benign masses, are usually asymptomatic until they reach a large size when they compress and displace adjacent structures. Moreover, several patients can present with constipation or pain due to local mass effect

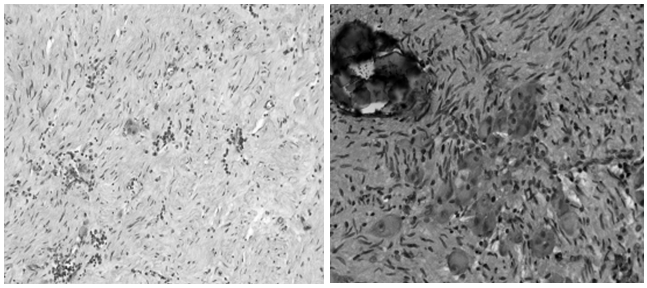

Figure 3 Hematoxylin and Eosin photomicrograph (× 10; × 20). Large ganglion cells are embedded in a neuromatous stroma with calcifications.

on the rectum, sacral root and lumbosacral plexus[1,23]. Ganglioneuromas have usually a mean diameter of 7 cm, so our patient is a rare case both for its presacral location and size[6]. On diagnostic evaluation, magnetic resonance represents the best non-invasive imaging method for preoperative study of this kind of lesions. Structural and morphological features of ganglioneuromas, such as the presence of mature sympathetic ganglion cells, can help distinguish them in differential diagnosis from other pelvic or abdominal lesions such as Schwannoma, neurofibroma, meningoma or cystic lesions. Lymphomas, chordomas, teratomas, soft-tissue sarcomas, Ewing sarcoma, osteosarcomas, chondrosarcomas or bone metastases are distinguished from ganglioneuromas because they are able to invade and erode bones. Preoperative study should be completed by FNAB samples of the lesion, but because of its anatomical location, technical approach is often difficult. Furthermore, an inaccurate diagnosis may occur with a single FNAB sample and multiple sites should be reached within the tumor[19]. So, we preferred intraoperative frozen section in order to define a correct diagnosis or discriminate between benign and malignant lesions. We preferred anterior laparotomic approach transperitoneally with rectum mobilization so as to achieve a complete tumor resection. A posterior trans-sacral approach was not preferred because of the potential iatrogenic morbidity to the dural sac and cauda equine. It was not necessary to perform laminectomy and foraminotomy because our patient was neurologically asymptomatic and intraoperative frozen section reported a benign lesion. To achieve this kind of surgical approach, we preserved nerve tissues avoiding morbidity and neurological dysfunction for the patient. If necessary, we could perform laminectomy and foraminotomy in case of recurrence with neurological symptoms. Because of the benign nature of ganglioneuromas, adjuvant systemic chemotherapy or local radiotherapy are not indicated. Moreover, surgical resection represents the only choice for both diagnosis and treatment, and a regular follow-up is necessary to assess local recurrence. Ganglioneuromas have a tendency to remain silent for a long time, and are often associated with a long-term disease-free survival[23]. Therefore, annual follow-up with neurologic examination and pelvic magnetic resonance is necessary, particularly when residual disease is present after surgical debulking.

Table 1 Clinical and surgical data in our patient and 17 previously reported cases

Authors	Age (yr)/Sex	Symptoms	Surgical approach	Resection
MacCarty (1965)	37/M	None	Transperitoneal approach + sacral amputation	C
Andersen (1986)	14/M	None	Transperitoneal approach	C
Richardson (1986)	71/M	Neurogenic bladder, constipation	Sacral laminectomy	P
Leeson and Hite (1989)	21/F	Pain, weight gain, urinary frequency	Transperitoneal approach	P
Stener (1989)	20/F	Pain	Sacral amputation	C
Spirnak and Wood (1993)	8/M	Constipation	Not specified	C
Okai (2001)	70/M	Constipation, pain	Transperitoneal approach	P
Lam and Nagib (2002)	11/M	Constipation	Transperitoneal approach + laminectomy	C
Marmor (2002)	70/M	None	Transperitoneal approach	C
Modha (2005)	65/F	Pain	Retroperitoneal exposure	P
	21/F	Pain	Retroperitoneal exposure	P
	21/M	None	Transperitoneal approach	P
	19/F	Constipation, low back pain	Transperitoneal approach	C
	28/F	Low back pain	Transperitoneal approach	P
Przkora (2005)	17/F	Amenorrhoea, weight loss	Sacral resection+laminectomy	C
Cerullo (2005)	64/M	None	Transperitoneal approach	C
Mounasamy (2006)	64/M	Low back and leg pain	Transperitoneal approach + laminectomy	C
	21/F	None	Transperitoneal approach + laminectomy	P

C: complete rection; P: partial resection; M: male; F: female.

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