Case Report

Pudendal Neuralgia as the Initial Manifestation of Infiltrative Sacrococcygeal Chordoma

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Keywords

Sacrococcygeal chordoma · Sacral tumours · Pudendal neuralgia

Abstract

Sacrococcygeal chordoma is a malignant tumour originating from remnants of the notochord. Chordomas are slow-growing tumours whose symptoms develop insidiously. We present the case of a 72-year-old woman with a 6-month history of genital pain radiating to the perianal area and exacerbating when she was in a sitting position. MRI and PET studies revealed a large mass in the sacrococcygeal region causing bone destruction and invasion of neurovascular structures. The immunohistochemical study of the surgical specimen determined it to be chordoma. This is the first published case of pudendal neuralgia as a form of presentation of sacrococcygeal chordoma.

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Introduction

Chordoma is a rare type of tumour (accounting for 0.2% of all central nervous system tumours and 1–4% of all primary bone tumours) originating from ectopic embryonic notochord remnants along the neuroaxis [1–4]. Its annual incidence is approximately 0.1 cases per 100,000 [4]. Chordomas typically develop in the sacrococcygeal and spheno-occipital

areas. The most common initial symptom of sacrococcygeal chordomas is local pain with sciatic radiation [2, 4].

Pudendal neuralgia is an extremely rare form of presentation of sacrococcygeal chordoma. Ours is the first case published in the literature.

Case Report

Our patient was a 72-year-old woman with no known drug allergies and a history of mixed tension migraine and essential tremor. The patient reported a 6-month history of pain in the genital area bilaterally which radiated to the perianal area and increased when she was in a sitting position. She was evaluated by multiple specialists and underwent rehabilitation therapy, with no relief. At that time, physical examination only revealed gluteal muscle contracture and pain upon palpation of the piriformis muscle. Pain did not subside and 4 months later she began to experience rectal tenesmus and urinary incontinence. The examination showed progression of neurological symptoms with saddle anaesthesia and left S1–S2 hypoaesthesia.

An MRI scan of the lumbosacral region revealed a giant multilobular soft-tissue mass measuring 90 × 73 × 57 mm and destroying the lower half of the left sacral ala and invading the left gluteal muscles and presacral space (fig. 1). This resulted in vessel displacement and infiltration of the presacral venous plexus at that level. A PET scan showed a hypermetabolic mass in the sacrococcygeal area; no abnormal uptake was seen in other areas. Tests for tumour markers α -fetoprotein, carcinoembryonic antigen, and CA 19–9 yielded negative results.

After complete surgical resection of the tumour, the patient progressed favourably. The histological study of the mass revealed cords of cells in a basophilic stroma. Epithelioid cells were polygonal in shape with eosinophilic cytoplasm, nuclear pleomorphism with hyperchromatism, and prominent nucleoli (physaliferous cells). Areas of congestion and haemorrhage were also seen. Immunophenotyping indicated that cells were positive for cytokeratin (AE1/AE3), S100 protein, epithelial membrane antigen, vimentin, and mesothelium (fig. 2, fig. 3).

Discussion

Sacrococcygeal chordoma is a rare entity that was first described by Rudolf Virchow in 1857 [2]. It is a slow-growing tumour arising from notochordal cells [3]. The notochord is a rod-like structure which develops in chordate embryos and constitutes the axis of the body. During fetal development, the notochord regresses, except at the location of the future intervertebral discs where it contributes to forming the nucleus pulposus. Notochord remnants can be found in adults at the base of the skull, the odontoid process of the axis, and the coccyx. This may explain why chordomas are more frequently seen at the 2 ends of the vertebral column [2]. They are malignant tumours that invade and destroy adjacent structures [5]. Chordomas are more common in men (male-to-female ratio of 2:1); mean age at presentation is 50–60 years [2, 6].

Symptoms are rare in early stages, with pain (lumbosacral pain or coccygodynia) being the first manifestation of the tumour [2, 7–9]. They are normally non-specific (pain, constipation), and diagnostic delays may therefore be considerable (range, 0.7–120 months) [10,

11]. On rare occasions, the tumour capsule may rupture and the tumour mass may invade the medullary cone, resulting in cauda equina syndrome secondary to compression or infiltration [12, 13].

In our patient, the initial manifestation of the tumour was persistent pain in the perineum over the course of 6 months, a finding compatible with pudendal neuralgia. The pudendal nerve originates from the ventral rami of sacral roots S2, S3, and S4 [14]. It extends under the sacrospinal ligament and through the pudendal canal below the levator ani muscle. It divides into 3 branches (inferior rectal nerve, perineal nerve, and the dorsal nerve of the penis/clitoris) which contain motor, sensory, and autonomic fibres [14]. To our knowledge, this is the first published case of pudendal neuralgia as the initial manifestation of sacrococcygeal chordoma.

In a follow-up consultation 4 months later, neurological symptoms were found to have progressed to cauda equina syndrome. The time elapsed from symptom onset to diagnosis was 10 months; this diagnostic delay, which falls within the average range, is due to the fact that symptoms are non-specific and appear insidiously.

MRI is the diagnostic method of choice; this technique enables accurate assessment of the lesion and aids in surgical planning. It is also the tool that best shows the extension of extraosseous infiltration and how it affects other structures. Lesions are hypo- or isointense on T1-weighted sequences and markedly hyperintense on T2-weighted sequences [5]. Images display hyperintense lobules separated by hypointense septations [5]. This pattern corresponds to the lobes and septations described in the histological study.

An anatomical pathology study provides the definitive diagnosis. From a macroscopic viewpoint, chordomas consist of friable masses of neoplastic tissue with a lobular pattern, a myxoid or gelatinous appearance, and a bluish-white or grey colour [6]. Tumour cells may display epithelioid features, with abundant eosinophilic cytoplasm (physaliferous cells) and immunohistochemistry positivity for keratin (predominantly cytokeratin 5), S100 protein, and epithelial membrane antigen [2, 6]. Positivity for S100 protein rules out carcinomas, which are negative for this marker, and positivity for cytokeratins rules out chondrosarcoma [1].

Radical surgical resection is the treatment of choice. Although it is not curative, high-dose radiotherapy may be beneficial when complete resection is not possible due to tumour infiltration into adjacent, non-resectable structures [2, 3].

Conclusion

Sacrococcygeal chordoma is a rare entity manifesting with non-specific symptoms for long periods of time (months or even years), which acts as an obstacle to early diagnosis. This type of tumour should be suspected in patients presenting lumbosacral or coccygeal pain associated with symptoms of spinal root or spinal cord compression.

Statement of Ethics

The authors have no ethical conflicts to declare.

Disclosure Statement

The authors report no conflicts of interest.

References

- 1 Torres Gómez FJ, Torres Olivera FJ, Torres Gómez A: Sacrococcygeal chordoma. Cyto-histological correlation. *Rev Esp Patol* 2008;41:293–296.
- 2 Walcott BP, Nahed BV, Mohyeldin A, Coumans JV, Kahle KT, Ferreira MJ: Chordoma: current concepts, management, and future directions. *Lancet Oncol* 2012;13:69–76.
- 3 Gronchi A, Casali PG, Olmi P: Sacro-coccygeal chordoma: diagnosis and treatment review. *Semin Colon Rectal Surg* 2004;15:26–32.
- 4 Ferraresi V, Nuzzo C, Zoccali C, Marandino F, Vidiri A, Salducca N, et al: Chordoma: clinical characteristics, management and prognosis of a case series of 25 patients. *BMC Cancer* 2010;28:10–22.
- 5 Farsad K, Kattapuram SV, Sacknoff R, Ono J, Nielsen GP: Sacral chordoma. *Radiographics* 2009;29:1525–1530.
- 6 McCormick M, Schroeder T, Benham S: Sacral chordoma: a case report with radiographic and histologic correlation and a review of the literature. *WMJ* 2006;105:53–56.
- 7 Blocker O, Hill S, Woodacre T: Persistent coccydynia – the importance of a differential diagnosis. *BMJ Case Rep* 2011, DOI: 10.1136/bcr.06.2011.4408.
- 8 Williams SP, Beckerman B, Piña Fonti ME: A 69-year-old presenting with musculoskeletal low back pain: a case of lumbosacral chordoma. *J Chiropr Med* 2014;13:144–148.
- 9 Gavrilidis P, Kyriakou D: Sacrococcygeal chordoma, a rare cause of coccygodynia. *Am J Case Rep* 2013;14:548–550.
- 10 Aydin R, Aydin G: An unusual cause of constipation: sacral chordoma. *Spine J* 2015;15:51–52.
- 11 Jeys L, Gibbins R, Evans G, Grimer R: Sacral chordoma: a diagnosis not to be sat on? *Int Orthop* 2008;32:269–272.
- 12 Ramdass S, Brennan M: A rare cause of cauda equina syndrome by infiltration: case of a sacral chordoma. *J Am Geriatr Soc* 2015;63:1269–1270.
- 13 Tharmabala M, LaBrash D, Kanthan R: Acute cauda equina syndrome secondary to lumbar chordoma: case report and literature review. *Spine J* 2013;13:35–43.
- 14 Khoder W, Hale D: Pudendal neuralgia. *Obstet Gynecol Clin North Am* 2014;41:443–452.

Fig. 1. MRI scan of the lumbosacral area. Non-contrast and gadolinium-enhanced T1-weighted sequences (a, c) and T2-weighted sequences (b).

Fig. 2. **a** Conventional panoramic view showing the lobular pattern of the tumour, which appears separated by septa composed of connective tissue of different thickness (H&E $\times 20$). **b** Border area of a lobe framed by a fibrous septum. The fine anastomosing cords of the tumour sample are cords immersed in a weakly basophilic myxoid matrix (H&E $\times 40$). **c** Detail of the previous figure. The cells show multivacuolated cytoplasm and some have two or more nuclei, hyperchromatic and slightly irregular, showing the characteristics of physaliferous cells (H&E). **d** The strands of the neoplastic cells appear anastomosing, showing an aspect of epithelioid cell characteristics (H&E).

Fig. 3. **a** The neoplastic cells are positive for cytokeratin immunostaining (AE1/AE3) ($\times 40$). **b** Focal positivity for S100. **c** Positivity for mesothelium. **d** Strongly positive for vimentin.