An unusual presentation of a rare tumor: Sacrococcygeal Chordoma presents with constipation

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An unusual presentation of a rare tumor: Sacrococcygeal Chordoma presents with constipation

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Abstract

Introduction: Chordoma is a rare, low grade and slow growing malignant tumor. The diagnosis of chordoma is often delayed due to non-typical signs and symptoms of disease. The most common symptoms are pain and neurological impairment. Case presentation: A 46 years old man presented with hypogastric abdominal pain and constipation for 4 months and evaluation revealed no abnormal findings. 7 months later he returned with low back and buttock pain with exacerbated constipation, fecal and urinary incontinency and weight loss. Physical examinations revealed a hard mass with small tenderness, approximately 15×20 cm in the sacrococcygeal region which was also palpated posterior to rectum in rectal examination. He went under surgery and tumor was excised. In pathology evaluation chordoma was diagnosed. Conclusion: Our case demonstrates an unusual presentation of chordoma. Due to different kinds of signs and symptoms in patients with chordoma only high clinical index of suspicion can help when a patient presented with constipation and may cause an earlier diagnosis, thus improving the prognosis.

KEYWORDS: Chordoma, Constipation, Sacrococcygeal
Introduction:

Chordoma is a rare, low grade and slow growing malignant tumor that originates from embryonic notochordal remnants. (1,2,4) It may appear across the midline of cerebrospinal axis; however, it occurs in the sacral area in males in 5th or 6th decade of life most frequently.(2,3) Chordoma has low tendency to distant metastasis, however the most common sites of metastasis are lungs and axial skeleton.(1,2,4) The diagnosis of chordoma is often delayed due to non-typical signs and symptoms of disease.(4) The most common symptoms are pain and neurological impairment. The pain often occurs in anal and lumbosacral region and also in lower back with radiation of pain to the legs.(1,4) The character of pain is typically dull (5) and neurologic deficits tend to be variable based on the location of lesion(6) Tumor size can reach more than 11 cm of maximum diameter(1) thus they are able to make signs and symptoms due to the compression of the mass to the neighboring organs. The case that is introduced in this article was presented with gastrointestinal complaints.

Case Report:

A 46 years old man was admitted in gastroenterology department due to hypogastric abdominal pain and constipation from 11 months ago. He had no other symptoms. In physical exam he had no abnormal sign and digital rectal examination was normal. Colonoscopy revealed no abnormality. He was discharged with medical therapy and advice for follow up. 7 months later he was presented with low back and buttocx pain with exacerbated constipation and weight loss. He had intermittent fecal and urinary incontinence from 2 months ago. The patient’s medical and family histories were unremarkable, only he had a Hemorrhoidectomy last year. On clinical examination, there was a hard mass with small tenderness, approximately 15×20 cm in the sacrococcygeal region with no change of color. (Fig.1). On rectal examination, a rubbery, mild tender and partially mobile mass was detected in pelvis, posterior to rectum without invading it (any invasion). Motor and sensory function of both lower limbs and anal reflexes were all normal. Other physical exams were normal except the bitemporal atrophy. Pelvic X-ray showed sacral destruction (Fig.2). In reconstructive MRI, an expansive and destructive lesion was seen in sacrum and coccyx (Fig.3). All preoperative laboratory studies were normal. He became candidate for sacrococcygeal excision with both transabdominal and posterior approach after mechanical and chemical colon preparation.
In the operating room, under general anesthesia, abdomen opened with midline incision below the umbilicus and rectum and ureters released. The tumor was released from pelvic organs and feeding vessels of the tumor were ligated. After closing the abdomen the patient was turned to the prone position and sacraococcygeal excision with preservation of S1-S3 nerve roots was done by our neurosurgeon colleagues.

In the pathology evaluation, the tumor was reported as conventional type of chordoma with extensive bone and soft tissue destruction. Eosinophilic and vacuolated cytoplasms with vesicular nucleus were seen in the cells. Pleomorphism and moderate atypia and lobular pattern were seen in myxoid stroma with necrotic focuses with invasion to bone trabecules.

The patient stayed in the ICU for one day and then he was transferred to the ward. He was ambulated on the 4th post operative day and was discharged on the 8th post operative day in good condition.

Discussion:

Chordoma is a rare tumor and diagnosis is usually delayed due to the number of symptoms that correlate with this pathology.\(^1\) Because of the wide range of differential diagnosis according to the symptoms, most of the patients undergo multiple investigations before diagnosis of chordoma.\(^2\) This long process may result in losing the time or missing the patients like this patient that was first presented with constipation and different work-up could not reveal the sacral mass possibly due to the small size of the tumor that time. Although chordoma is a slow growing tumor\(^2\), after 7 months in our patient the mass was detectable in physical and rectal exam and the symptoms were developed to low back and buttock pain, chronic constipation and episodes of fecal and urine incontinency. The only definite treatment in chordoma is radical surgical excision of the tumor.\(^3\) Early diagnosis may lead to more easy resection due to the smaller size of the tumor and less subsequent morbidities.\(^4\) Excision of the larger tumor may necessitate the excision of the sacral nerve roots that results in sexual dysfunction, permanent incontinence and motor weakness.\(^5\) Thus for early diagnosis that may lead to the curative surgery, in patients with bowel or urinary disturbances even without low back pain and paresthesia or motor weakness in lower limb, sacral tumor especially chordoma should be considered as one of the differential diagnosis.
References:

