Extraosseous chordomas are extremely rare. In this case report, we describe an extraosseous chordoma in the coccygeal region that later metastasized to the cerebellum. A 30-year-old man presented with a progressively enlarged lesion in the coccygeal region with painful sensation. Preoperative computed tomography and magnetic resonance imaging showed a large lobular mass in the coccygeal region without sacrococcygeal destruction. Pathology showed physaliphorous cells and the immunohistochemical staining pattern typical of chordoma.

Key words: Computed tomography; Extraosseous chordoma; Magnetic resonance imaging

Chordomas are uncommon malignant tumors of notochordal origin that arise along the craniococcygeal axis. The distribution of chordomas is about 50% in the sacrococcygeal region, 35% in the clivus, and 15% in the vertebrae [1]. The peak incidence of chordoma is in the fifth and sixth decades of life, with a male predominance of about 2:1 in many series [2]. Extraosseous chordoma is very rare [3]. Soft-tissue chordoma without skeletal involvement has been rarely reported, with most cases being described in the mediastinum [4].

Sacrococcygeal chordomas often present as lower back pain, with pain as the earliest and most common symptom. Classic chordomas are usually locally invasive, with low metastatic potential [5]. Both hematogenous and lymphatic metastatic pathways have been reported. The most common metastatic site is the lung [6], whereas cerebellar metastasis are rare [7]. The pathological and immunohistological characteristics of chordomas are very distinctive in that they contain physaliphorous cells with positive immunoreactivity for cytokeratins of different molecular weights, vimentin, S-100 protein, and epithelial membrane antigen (EMA). We present one case of extraosseous chordomas with distant metastases to the cerebellum and lung.

CASE REPORT

A 30-year-old man presented with recent bowel habit changes associated with a painful sensation. He had experienced a slowly enlarging mass with discharge, located over the coccygeal region for the previous two years.

Physical examinations were generally normal, except for a reddish and protruding mass over the coccygeal region. Routine complete blood count results and his serum biochemical profile were unremarkable. A pelvic computed tomography (CT) scan (Fig. 1) showed a lobular heterogeneous mass...
of more than 22 cm over the coccygeal region with coccygeal encasement and no coccygeal destruction. The rectoischial space was completely replaced by the tumor. A CT demonstrated a tumor in contact with the posterior rectal wall. A barium enema was administered, which showed a normal mucosal pattern in the colon and poor distention of the rectum owing to an extrinsic posterior mass compression. Cystoscopy showed no tumor invasion into the urinary bladder. Pelvic magnetic resonance imaging (MRI) (Fig. 2) was performed using a 1.5 tesla MR scanner (Siemens Vision Plus, Erlangen, Germany). A huge mass was identified in the caudal aspect of the coccygeal region, which involved the subcutaneous soft tissue in the gluteal region posteriorly, and displaced the rectum and urinary bladder anteriorly. The mass showed an intermediate signal intensity with heterogeneous enhancement on spin-echo T1-weighted images (TR/TE/excitations, 144/4.1/1) (Fig. 2a) after gadolinium administration and heterogeneous hyperintensity on spin-echo T2-weighted images (TR/TE/excitations, 4400/90/1) (Fig. 2b). The fat plane between the posterior wall of the rectum and the tumor was not clear. Because the tumor extended to the perianal region and the right-sided gluteus musculature at surgery, both exploratory laparotomy with the removal of the tumor and colostomy were performed. Postoperative adjuvant radiotherapy and chemotherapy was arranged. Gross pathology revealed a mass measuring 25 cm ~ 30 cm, located in the caudal aspect of the coccygeal region, gray-black in color with a lobular contour, and with hemorrhage and extensive necrosis. No calcified component within the mass or bony destruction of the coccygeal region was noted.
Microscopically (Fig. 3), the specimen revealed a pattern of physaliphorous cells in a mucinous background, the appearance typical of a chordoma. The neoplastic cells were positive for cytokeratin, S-100, EMA, and vimentin. All these features reconfirmed the initial diagnosis of chordoma [7].

Intrapulmonary metastases developed two months later. A CT scan (Fig. 4a) was performed one year later, which showed a soft-tissue nodule, about 1.5 cm in diameter, in the left upper lung. A thoracotomy with wedge resection was performed. Chordoma metastasis to the lung was confirmed by histopathology.

Dizziness and an unstable gait developed three years later. A brain MRI (Fig. 4b) was performed and showed a complex mass lesion about 3.8 cm in size, with a heterogeneously enhanced nodule and cystic component, in the right cerebellar hemisphere. Extraosseous chordoma with brain metastasis was confirmed by histopathology.

The patient received postoperative adjuvant radiotherapy and several courses of chemotherapy. Enlarged nodes in the mediastinum and a newly developed nodule of about 0.8 cm in the left upper lung were identified five years later. The patient is currently stable and no local recurrence has been identified in the coccygeal region after six years.
DISCUSSION

The classic chordoma is a slow-growing, local, aggressive neoplasm and uncommon malignancy derived from the embryonic remnants of the notochord along the craniococcygeal axis, mostly occurring in the sacrococcygeal region. The notochord structure is associated with the development of the axial skeleton, appearing in the fourth week of gestation and regressing by the seventh week. Incomplete involution of the notochord results in chordoma [2]. In contrast, the rare extraosseous chordoma is believed to have its origins in ectopic notochordal rests separated from the bone, or originating from a clinically undetectable primary bone tumor with growth into the soft tissue [5].

According to a new classification of spinal tumors, chordomas are described as follows: type I, intraosseous extradural; type II, intraosseous intradural; type III, extraosseous extradural; type IV, extraosseous intradural; and type V, extraosseous soft tissue [2]. We consider our case to be a type V chordoma. The locations of unusual extraosseous chordomas, such as the tentorium cerebelli, terminal filum, intradural foramen magnum, intradural cervical regions, intradural thoracic regions, and epidural lumbar regions, have been reported in the literature [8]. Among the extraosseous chordomas reported in the literature, 15 were extraosseous intradural, three were extraosseous extradural, and one was extraosseous soft tissue [2]. Two unusual cases of extraosseous chordomas of the sacrococcygeal region have also been reported in the literature [5, 7].

The metastatic potentials of the extraosseous and classic chordomas have been compared. Classic chordomas are locally invasive neoplasms that may metastasize to distant sites [9]. Our review of the literature did not reveal any case of intradural extraosseous chordoma with a potential for distant metastasis because the removal of the intradural extraosseous chordoma was complete [10]. Complete resection of an intradural extraosseous chordoma is feasible because of their sharply circumscribed margins. In contrast, classic chordomas are not easily resectable because of osseous involvement and their ill-defined margins [10]. About 76% of classic chordomas that metastasize do so as a result of incomplete excision. Incomplete excision may allow the residual tumor to gain access to the bloodstream and metastasize distantly [9]. The metastatic locations of classic chordomas are, in decreasing order of frequency, the lungs, liver, skeleton, lymph nodes, skin, subcutaneous regions, spleen, adrenal glands, kidney, urinary bladder, and pancreas [6, 11-13]. Brain metastasis is rare, with only seven cases of chordomas that metastasized to the brain reported in the literature [9].

Many researchers have tried to identify prognostic factors for chordoma. Bergh et al. [14] found that an inadequate surgical margin at initial surgery and the presence of microscopic tumor necroses are adverse factors for chordoma. Cheng et al. [13] found that a better prognosis correlates with a more proximal position. Sell et al. [15] considered that the recurrence of chordoma is related to nuclear atypia and pleomorphism but not to the Ki-67 index. Radiologically, several authors have also suggested that T1 and T2 factors help to distinguish benign and malignant tissues in vivo. However, recent evidence suggests that these indicators are not specific [16].

A correlation between pathological findings and imaging features has been reported in the literature. Both increased water and fluid content and the presence of proteinaceous and mucinous material within the matrix are possible factors on hyperintense T2-weighted MR images [17]. These fibrous septa of the tumor appear as thin linear bands of low signal intensity on T2-weighted MR images. High-signal foci on T1-weighted MR images are due to intratumoral hemorrhage [17].

Chordomas that are entirely extraosseous are rare [10]. The vast majority of chordomas have characteristic bone destruction. The diagnosis in our patient was supported by the location of the growth and the gross and pathological findings. The images of this sacrococcygeal chordoma are characteristic because there is no sacrococcygeal destruction. In our patient, the mass showed heterogeneous enhancement on T1-weighted MR images and heterogeneous hyperintensity on T2-weighted MR images. Signal intensity in MRI studies is not specific for a diagnosis of chordoma because the MR images are similar for many tumors.

Radiologically, sacrococcygeal chordoma must be differentiated from primary osseous tumors and teratomas, and neurogenic tumors such as sacral schwannoma, metastatic neoplasm, soft-tissue malignant fibrous histiocytoma, and rectal leiomyosarcomas [18, 19].

Metastatic carcinomas are the most common sacral neoplasms, with lung, breast, kidney, and prostate carcinomas the most frequent causes. Therefore, metastatic lesions should be considered first [18]. There was no evidence of a primary tumor...
in our patient, so a diagnosis of a metastatic lesion to the coccygeal region was less likely.

Osseous tumors, such as giant cell tumor, chondrosarcoma, aneurysm bone cyst, osteochondroma, lymphoma, Ewing sarcoma, fibrosarcoma, and osteosarcoma of the sacrococcyx with soft-tissue invasion in the retrorectal space, comprise approximately 5% to 10% of all retrorectal tumors [18]. Osseous tumors should be taken into consideration. However, the final diagnosis must be confirmed by pathological diagnosis.

One of the differential diagnoses of sacrococcygeal extraosseous soft-tissue tumor is rectal leiomyosarcoma, which is a malignant tumor of the smooth muscle. The tumor may involve the rectal mucosa, causing surface ulceration and bleeding. On CT, it may show a well-circumscribed, heterogeneously enhanced, multilobular tumor with cystic necrotic components [20]. In our patient, no evidence of mucosal ulceration of the rectum was identified with a barium enema study, so a diagnosis of rectal leiomyosarcomas was less likely based solely on the base of imaging studies.

Primary soft-tissue malignant fibrous histiocytoma usually appears as a nonspecific soft-tissue mass. Generally, it presents as an intermediate signal intensity on T1-weighted MR images and heterogeneous high signal intensity on T2-weighted MR images [21]. In our patient, the chordoma showed isointensity on T1-weighted MR images and heterogeneous hyperintensity on T2-weighted MR images. Because of its similar MRI appearance, soft-tissue malignant fibrous histiocytoma should be considered.

Another important differential diagnosis of sacrococcygeal tumors is sacrococcygeal teratoma. Generally, a sacrococcygeal teratoma contains mixed calcification, cystic components, fat–fluid debris levels, bone, hair, and cartilage [19]. In our patient, these typical components were not seen on the imaging study.

Sacral schwannoma and neurofibroma that occur in the sacrococcygeal region arise from the lower lumbar and sacral dorsal sensory-nerve roots. They may grow along the nerve segment and expand into the sacral canal and neural foramen [19]. No neural foraminal expansion was noted in our patient, so a diagnosis of sacral schwannoma or neurofibroma was less likely.

Although radiological images can help us differentiate the possible diagnoses of sacrococcygeal lesion, surgical resection and histological evaluation allow an absolute diagnosis of sacrococcygeal chordoma. The characteristic pathological feature is vacuolated cells that contain intracytoplasmic mucus, which are called physaliphorous [22]. Immunohistochemical features are also characteristic, in that chordomas are positively immunoreactive for cytokeratins of different molecular weights, vimentin, S-100 protein, and EMA [7]. The correlation of hematoxylin–eosin staining with the classical immunohistochemical profile of the chordoma was important in our patient and led to the correct diagnosis.

In summary, we have presented an unusual case of extraosseous chordoma in the coccygeal region with rare distant metastases to the cerebellum and lung. Chordoma should be considered in the differential diagnosis of a mass in the sacrococcygeal region, whether or not bony destruction is present.

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未侵犯薦尾骨的薦尾區脊索瘤合併小腦轉移：
病例報告

廖見銘  高鴻文  薛俊仁  阮春榮  羅中平  張維洲  陳震宇

國防醫學院  三軍總醫院  放射診斷部

非源自骨頭的脊索瘤臨床很少見，這個報告中，我們描述一個未侵犯薦尾骨的薦尾區脊索瘤病例，並且和併有罕見的小腦轉移，這位年輕病例過去兩年中發現薦部有腫塊且且逐漸長大併發疼痛感加劇，電腦斷層及磁振造影顯示出薦尾骨有個大的軟組織腫瘤，而且腫瘤並無侵犯薦尾骨，經術後病理診斷為脊索瘤。

關鍵詞：電脳斷層；薦尾脊索瘤；磁振造影