CASE REPORT

Malignant Clival Chordoma with Postoperative Cutaneous Metastases

William T. Couldwell, M.D., Ph.D.,
Charles B. Stillerman, M.D., Dale Rice, M.D.,
Dennis Maceri, M.D., Randolph Sherman, M.D.,
Takanori Fukushima, M.D., and David R. Hinton, M.D.

Abstract—The authors report the case of a chordoma with malignant cytologic features, presenting as a mass lesion in the clival and infratemporal region at the level of the cranio-cervical junction in an 8-year-old female. Following gross resection of the mass, the patient subsequently developed distant subcutaneous and peritoneal metastases from the lesion. The rare histologic features, the surgical approach to the lesion, and the follow-up management of this unique case are discussed. (*Skull Base Surgery*, 6(1):61–66, 1996)

Chordomas and chondrosarcomas are distinctly uncommon tumors, constituting less than 1% of intracranial neoplasms.^{1–3} Chordoma has been considered as a primary adult tumor, characterized by local invasive behavior with bone destruction and soft tissue infiltration, but slow overall growth.

We report here a case of a malignant chordoma in an 8-year-old Caucasian female which demonstrated aggressive histologic features and a rampant clinical course with postoperative distant cutaneous and peritoneal metastases.

CASE REPORT

This previously healthy Iranian female first complained to her parents about intermittent headache and neck pain at the age of five. The neck pain limited head motion when turning to the left, and as a result the patient held her head in a persistent right lateral position. Some months later she developed nausea and vomiting which prompted a visit to a neurologist in her native country. Computed tomographic examination at that time revealed a partially cystic lesion which, enhanced with intravenous contrast, was seen to be primarily extracranial with exten-

sion into the caudal clivus, occipital condyle, and posterior fossa on the left. As the patient was developing increasing throat pain with swallowing, as well as gait instability, she underwent a subtotal resection of the tumor via a suboccipital craniectomy in western Europe. Pathologic examination of the resected tumor at that time was interpreted to be that of fibrous histiocytoma, and she underwent a course of postoperative local radiotherapy (55 cGy). Her family had recently immigrated to the Los Angeles area, so she was subsequently referred to our institution for further oncologic and neurosurgical management. The patient was placed on a chemotherapeutic protocol (etoposide, ifosfamide, carboplatinum, and mesna, Q 3 weeks) with limited response; neurosurgical consultation was then obtained approximately 1 year following her initial surgery.

Examination

Physical examination was remarkable for a well-healed surgical incision and alopecia in the left suboccipital region, and left-sided tongue weakness and atrophy. Gait and cerebellar testing was within normal limits. In the year that had elapsed between the previous surgery

Figure 1. Preoperative axial (A) and sagittal (B) gadolinium-enhanced MRI images revealing a tumor mass located in the region of the clivus and the retropharyngeal soft tissue of the oropharynx on the left. Note the erosion of the clivus and occipital condyle on the left, with extension of the abnormal signal into the posterior fossa. The vertebral artery is shown to be encased by the tumor on the left.

and her evaluation at our institution, MRI examination revealed recurrence of the tumor in the clival and pre-clival region at the level of the cervical-medullary junction (Fig. 1).

Surgical Management

An extreme lateral transcondylar exposure approach was chosen for the second operation. Most of the tumor was anticipated to be primarily extradural, with some intradural component in this region based on the pre-operative imaging studies (Fig. 1). This approach allowed exposure to the tumor which had remained following the initial resection. It enabled both intradural and extradural access to the lateral skull base and the posterior fossa, as well as the retropharyngeal soft tissue space. At surgery, the tumor had involved the entire region of the occipital condyle, anterior C1, and the lower clivus. As the entire region of the lateral mass of C1 and the condyle was resected with the tumor (Fig. 2), she underwent a primary occipital-cervical fusion. The patient underwent an uncomplicated course, with gross total resection of the mass. All soft tissue margins biopsied were free of tumor at the time of the resection.

Twenty months following the resection, the child developed increasing neck pain, mostly at night and with attempted motion. Repeat MRI study revealed local recurrence of the tumor mass. She underwent a repeat transcondylar exposure, this time with accompanying infra-temporal exposure for removal of the recurrent tumor

Figure 2. Postoperative computed tomographic scan shows extensive basal bony removal. The arch of C1 and partial resection of the Dens (not shown on this image) has been accomplished with gross total resection of the tumor. The tumor bed is shown filled with autologous fat to obliterate dead space.

mass. At surgery, the entire inferior two thirds of the clivus and remainder of the C1 anterior arch were resected, with the large mass extending forward to the infratemporal fossa. All dura involved with tumor, and soft tissue involvement of the infratemporal fossa, was resected. The left vertebral artery was sacrificed and resected, extending from the foramen transversarium of C2 to the junction of the posterior-inferior cerebellar artery (PICA) intracranially. As much soft tissue was resected, a trapezius myocutaneous flap was transposed to the surgical defect for tissue coverage and CSF leak prophylaxis.⁴

Tumor Histology

The tumor had the histologic appearance of a chordoma with focal malignant cytologic features. The tumor widely infiltrated soft tissues and was densely cellular (Fig. 3A) with large areas of confluent necrosis. Mitotic figures were easily identified. The tumor cell nuclei showed focal, marked pleomorphism (Fig. 3B), and nucleoli were prominent. The tumor cells were discohesive on smear (Fig. 3C), and on section had an epithelioid morphology with abundant cytoplasm and prominent cell

Figure 3. Histologic appearance of the resected tumor. A: the cellular tumor is shown to be composed of epithelioid cells with prominent cell borders. Mitotic figures were easily identified (arrow). Many of the cell nuclei had prominent nucleoli. B: there was prominent nuclear pleomorphism in several areas. C: the discohesiveness of the tumor cells and the vacuolization of the cytoplasm (arrow heads) is shown on a smear preparation. Hematoxylin and eosin stain. Magnification $\times 110$ (A,B) $\times 400$ (C).

Figure 4. Cutaneous metastatic tumor spread. Note the biopsy proven extensive tumor deposits adjacent to the harvest site of the trapezius myocutaneous flap used to close the skull base defect.

borders (Fig. 3A). The cytoplasm was often vacuolated (Fig. 3C), although the classic physaliferous cells were not seen. Immunoperoxidase staining revealed that the tumor cells were strongly positive for cytokeratin, epithelial membrane antigen, and S-100. The tumor cells were negative for the premelanosome-specific antibody HMB-45.

Postoperative Course

The patient's immediate postoperative course was uneventful, with the exception of a new left vocal cord paralysis and associated hoarseness. Four months later, she developed signs and symptoms of raised intracranial pressure, which required placement of a ventriculoperitoneal shunt. Cerebrospinal fluid (CSF) examination was remarkable for atypical cells consistent with malignant cells. Four months following insertion of the shunt, she presented with abdominal pain and subcutaneous nodules along the dorsal harvest site of the trapezius myocutaneous flap (Fig. 4). Histologic biopsy of the cutaneous lesions revealed neoplasm consistent with the previous surgical specimens. Abdominal computed tomography (CT) revealed evidence of peritoneal masses compatible with metastatic tumor (not shown).

very rare in childhood, display a more variable and aggressive course with rapid growth, and more frequent metastases than these tumors in adults.⁷⁻¹⁰ Among 12 childhood chordomas reported in a recent review by Coffin et al,⁷ only one child or adolescent was alive and free of tumor 3 years after the initial diagnosis. In children, chordomas occur predominantly in the head and neck region, with the sacrococcygeal region being involved infrequently.⁷ Specifically, chordomas that arise in the first 2 decades of life have a predilection for the sphenoccipital and cervical regions.^{7,10-12} The present case supports the observations of the previously reported cases of chordoma in childhood,^{7,8,12-18} in that it demonstrated a markedly aggressive clinical course uncharacteristic of chordomas in adults.

Metastasis with Chordoma

There is a discrepancy in the literature as to the incidence of metastasis in patients with chordoma (ranging from 3%–48% of cases).^{11,19,20} The tumor can involve the skin by direct extension, local recurrence, or metastasis.²⁰⁻²² Locations of distant metastases in other reported childhood cases were to lungs, lymph nodes, and widespread.^{10,13,23,24} The present case is unique in that rampant metastases developed in cutaneous and peritoneal locations following resective surgery and CSF diversion, respectively. The presence of tumor cells in the spinal fluid at the time of insertion of the ventriculoperitoneal shunt and the later development of peritoneal metastases underline the importance of potential iatrogenic spread of tumor. Furthermore, the development of metastases in the region of the previous harvest site of the myocutaneous flap (Fig. 4) suggests direct seeding at

DISCUSSION

Chordoma in Childhood

Chordomas are tumors believed to derive from remnants of the embryonic notochord along the spinal axis.^{5,6} It has recently been suggested that chordomas, although

surgery or hematogenous-borne metastatic deposits which preferentially seed to regions of healing tissue. To our knowledge, the metastatic spread of a chordoma following surgical resection noted in the present case has not been reported previously. In addition, the presence of skin metastases from chordoma in the thoracic location as noted in the present case is distinctly unusual; in 19 of 207 patients (9%) with chordoma published by Su et al²⁰ with cutaneous involvement (coined *chordoma cutis* by these authors), all metastases were located only in the sacral region, scalp, face, or neck. None of these reported cases involved children.

Surgical Approach

In recent years, the advent of extended approaches to the skull base have facilitated radical resection of tumors in these locations. The extreme lateral approach to the occipital condylar region was first described by Seeger in 1978.²⁵ Subsequently, this approach has been popularized and refined by others.^{26–32} The approach offers excellent anterolateral exposure of the lower clivus and upper cervical spine, and is especially useful for access to lesions located anterior to the spinomedullary junction.

As these tumors are primarily extradural in location, Sen and Sekhar²⁷ make a valid point at directing a primary extradural surgical approach. The extreme lateral transcondylar exposure proved suitable for this purpose in that it enabled a wide exposure of the lateral posterior fossa skull base and upper cervical spine. At the third operation, the transcondylar approach was combined with an infratemporal exposure to remove the anteriorly-situated recurrent tumor.

Adjuvant Radio- and Chemotherapy

Conventional radiotherapeutic techniques have been relatively ineffective in controlling these neoplasms.^{33,34} The clinical course of the patient in the present report attests to the limited efficacy of external beam radiotherapy for tumors of this histology. Some authors have questioned the value of aggressive surgical resections in the hope of curing such an invasive lesion in this location, and have advocated improved contemporary methods of delivering local radiotherapy in the management of these tumors.^{35,36} However, the long-term control of focused and/or fractionated radiotherapy in the management of malignant chordoma, as in the present case, remains to be defined.

The patient in the present report achieved some limited local control of disease on combinational etoposide, ifosfamide, carboplatinum, and mesna therapy. Previously published experience with chemotherapy for primary and metastatic chordoma has been disappointing, and identification of further effective single or combina-

tional chemotherapeutic agents will be necessary to produce improvements in therapy for chordomas occurring in children.¹³

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REVIEWER'S COMMENTS

This case report adds further support to the premise that chordomas in children are more aggressive than those seen in adults and are often associated with metastases.

Leon Barnes, M.D.