Case Report

Parachordoma of Soft Tissues of the Arm

Jonathan Clabeaux MD, Leonard Hojnowski MD,
Alfredo Valente MD, Timothy A. Damron MD

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Abstract Parachordoma, or myoepithelioma, is a very rare tumor histologically resembling chordoma but occurring in the nonaxial soft tissues. It typically has an indolent nature, with occasional late recurrence and even rare metastases. Review of existing literature reveals a male predilection, with the tumor typically occurring in the fourth decade of life in the lower extremity. It typically is managed with wide resection. We report the case of a 60-year-old woman with a right distal upper arm parachordoma treated with wide resection of the tumor.

Introduction

Parachordoma, also known as myoepithelioma, is an extremely rare soft tissue tumor that histologically resembles chordoma of the axial skeleton in a nonaxial location.

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J. Clabeaux, T. A. Damron (✉)
Department of Orthopedic Surgery, Upstate Medical University,
Suite #130, 550 Harrison Street, Syracuse, NY 13202, USA
e-mail: damront@upstate.edu

L. Hojnowski
Department of Radiology, Upstate Medical University, Syracuse,
NY, USA

A. Valente
Department of Pathology, Upstate Medical University, Syracuse,
NY, USA

Fewer than 50 cases are reported in the English literature [14]. The first description is credited to Laskowski in 1951 [4], who initially labeled it chordoma periphericum. Dabska [4] later renamed it parachordoma and published the largest initial case series of 10 cases in 1977. Parachordoma is a soft tissue tumor characterized by an indolent nature, slow growth, occasional late recurrence, rare metastases, and at least two reported fatalities. Histologically, it resembles chordoma with a wider variation in appearance. We report the current case to increase awareness of this unusual soft tissue tumor.

Case Report

A 60-year-old right-hand-dominant woman presented with a soft tissue mass in the posteromedial aspect of her right distal arm of 3 months' duration. She denied any trauma to that area but stated the mass had slowly enlarged. She denied any pain, numbness, or tingling in the distal extremity. She also denied fevers, chills, weight loss, loss of energy, and loss of appetite but did have some occasional night sweats. She had a history of localized thyroid cancer in 1989 treated with thyroidectomy. She denied any lumps or bumps elsewhere.

Physical examination revealed a healthy-appearing woman with a palpable, firm, deep, slightly warm mass over the posteromedial aspect of her distal right arm that measured approximately 7 cm proximodistal by 4 cm mediolateral. Tinel's test was negative, and there was no bruit, thrill, or tenderness to palpation over the mass.

The initial radiographic studies included plain radiographs and MRI of the extremity. The radiographs showed a slight soft tissue density at the posteromedial aspect of the arm without any evidence of periosteal reaction or

calcification in the mass (Fig. 1). Magnetic resonance imaging revealed a complex, heterogeneous, deeply situated intramuscular soft tissue mass adjacent and posterior to the neurovascular bundle on the medial aspect of the arm in the triceps muscle belly. The mass was predominantly dark with heterogeneity on the T1-weighted images (Fig. 2A) and bright with some heterogeneity on T2-weighted images (Fig. 2B). There was minimal soft tissue edema surrounding the mass. There was no bony involvement and no periosteal reaction, but it did appear to abut the humerus medially.

The nonspecific MRI features of the deep, large heterogeneous soft tissue mass yielded a broad differential diagnosis, including benign and malignant entities such as malignant fibrous histiocytoma, fibrosarcoma, and even metastatic disease. Tissue for pathologic examination was obtained through a Tru-Cut core needle (Travenol Laboratories, Inc, Deerfield, IL) biopsy in the office. Staging studies, which included computed tomographic scans of the chest, abdomen, and pelvis, were negative for the presence of metastases. Laboratory values were all within normal

Fig. 1 A plain radiograph of the elbow and distal arm shows only a nonmineralized soft tissue shadow.

limits, including a leukocyte count of 6000, erythrocyte sedimentation rate of 15 mm/hour, and C-reactive protein less than 0.5.

The Tru-Cut needle biopsy specimen revealed a tumor composed of nests and cords of cells with eosinophilic and vacuolated cytoplasm and relatively uniform round to oval nuclei. The cells were embedded in a myxoid and hyaline stroma and separated by broad fibrous septa (Fig. 3). Foci of recent and remote hemorrhage were present, but necrosis was not identified. In addition, there was a low mitotic rate not exceeding one mitosis per 20 high-power fields (Fig. 4).

Differential diagnosis for these histologic findings included parachordoma, extraskeletal myxoid chondrosarcoma, and chordoma. Immunohistochemistry revealed expression of the cytokeratin CAM5.2 and S-100 protein by tumor cells with negative staining for cytokeratins AE1/AE3, cytokeratin 19, epithelial membrane antigen (EMA), muscle-specific actin, desmin, and carcinoembryonic antigen (CEA). An alcian blue stain was strongly positive in the stroma, with marked reduction of the staining after hyaluronidase digestion. The diffuse CAM5.2 and S-100 expression is an immunophenotype not seen in extraskeletal myxoid chondrosarcoma, although negative cytokeratin 19 and CEA with hyaluronidase digestion of the matrix favored parachordoma rather than chordoma. Because extraskeletal myxoid chondrosarcomas are characterized by the balanced chromosomal translocation $t(9;22)(q22;q12)$, with the breakpoint involving the *EWS* gene on chromosome 22q12 and the *CHN* gene on 9q22, fluorescence in situ hybridization studies for the detection of the *EWSR1* gene break-apart rearrangement were performed, and the results were negative.

The patient underwent wide resection of the tumor 2 weeks later. A curvilinear incision was made on the posteromedial aspect of the arm, which included excision of the biopsy tract. The ulnar nerve was identified and protected during the entire procedure. The mass was removed en bloc with negative intraoperative frozen section margins. She experienced some mild numbness along her small finger and the ulnar border of the ring finger but full abduction strength. The final pathologic results also were consistent with parachordoma. There has been no recurrence to date during the past 3 months of followup.

Discussion

Parachordoma (or myoepithelioma) is a rare soft tissue tumor first described by Laskowski and then renamed by Dabska [4] in 1977. The differential diagnosis often includes extraskeletal myxoid chondrosarcoma, and this was considered in the pathologic differential diagnosis in

Fig. 2A–B Sagittal MRI of the distal upper arm soft tissue mass shows a (A) heterogeneous dark signal on the T1-weighted image and a (B) heterogeneous bright signal on the T2-weighted image.

Fig. 3 A low-power photomicrograph of the tumor shows multinodular masses separated by broad collagen bands (Stain, hematoxylin and eosin; original magnification, $\times 20$).

Fig. 4 A high-power magnification with tumor cells shows vacuolated cytoplasm and lack of considerable nuclear atypia (Stain, hematoxylin and eosin; original magnification, $\times 400$).

the current case [8]. Parachordoma originally was believed to be a chordoma occurring in nonaxial sites, but more recent work suggests it has a distinct immunohistochemical profile [6]. Presently, parachordoma is considered a unique entity [14].

Light microscopic features of parachordoma include cells in clusters, nodules, whorls, and a pseudoglandular formation of rounded cells in a focally myxoid stroma separated by fibrous tissue [6]. In general, nuclei are bland, mitotic figures are rare, and there is no necrosis or vascular invasion. The tumor is circumscribed but not encapsulated.

Parachordoma is composed of three cell types, specifically epithelioid cells, smaller glomoid cells, and spindle cells [8]. All lesions have a population of cells with vacuolated cytoplasm resembling the physaliferous cells found in chordomas. Parachordoma and chordoma differ further in their expression of immunophenotypes. Both tumors show immunohistochemical evidence of positivity with CAM5.2 (which recognizes cytokeratins 8/18), EMA, vimentin, and S-100. However, reactivity to Type IV collagen is much stronger in parachordoma, whereas chordoma expresses CK1/10 and CK19 most of

Table 1. Clinical findings of parachordoma case reports

Authors (Year)	Age (years)	Gender	Site	Recurrence	Metastasis	Fatal
Dabska [4] (1977)	47	Female	Ankle	7 years	No	No
	52	Male	Knee	No	No	No
	18	Male	Thigh	No	No	No
	12	Male	Arm	No	No	No
	29	Female	Back	12 years	No	No
	50	Female	Groin	No	No	No
	22	Female	Forearm	No	No	No
	25	Male	Femur	Unknown	Unknown	No
	28	Female	Finger	2 years	No	No
	62	Male	Ilium	No	No	No
Povysil and Matejovsky [19] (1985)	Unknown	Unknown	Tibia	Unknown	Unknown	No
Enzinger and Weiss [5] (1988)	53	Male	Back	Unknown	Unknown	No
Miettinen et al. [17] (1992)	67	Female	Popliteal	No	Metastasis	Fatal
Ishida et al. [12] (1993)	19	Female	Calf	1 year	No	No
Shin et al. [22] (1994)	4	Male	Unknown	Unknown	Unknown	No
	8	Male	Unknown	Unknown	Unknown	No
	12	Male	Unknown	Unknown	Unknown	No
	13	Male	Knee	Unknown	Unknown	No
	15	Male	Unknown	Unknown	Unknown	No
	20	Female	Calf	Unknown	Unknown	No
Hirokawa et al. [10] (1994)	43	Male	Buttock	No	No	No
Sanguenza and White [20] (1994)	25	Male	Finger	Unknown	Unknown	No
Carstens [3] (1995)	42	Male	Buttock	6 months	Metastasis	Fatal
Niezabitowski et al. [18] (1995)	45	Female	Palm	3 months	No	No
Carlen [2] (1996)	45	Female	Leg	Unknown	Unknown	No
Karabela et al. [13] (1996)	86	Female	Pelvis	No	No	No
Fisher and Miettinen [7] (1997)	23	Male	Thigh	Unknown	Unknown	No
	25	Female	Triceps	No	No	No
	53	Male	Back	No	No	No
	14	Male	Wrist	Unknown	Unknown	No
	52	Female	Chest	2 years	Metastasis	Fatal
Imlay et al. [11] (1998)	13	Male	Thigh	No	No	No
	25	Female	Forearm	Yes	No	No
Tihy et al. [23] (1998)	7	Female	Nares	No	No	No
Folpe et al. [8] (1999)	42	Female	Forearm	No	No	No
	7	Female	Buttock	Unknown	Unknown	No
	45	Male	Chest	No	No	No
	38	Male	Deltoid	No	No	No
	30	Female	Thigh	No	No	No
	62	Male	Thigh	No	No	No
	64	Male	Tibia	Recurrence	No	No
Koh et al. [15] (2000)	20	Female	Hand	No	No	No
Separovic et al. [21] (2001)	24	Male	Knee	No	No	No
Hemalatha et al. [9] (2003)	68	Male	Calf	No	Metastasis	Fatal
Abe et al. [1] (2003)	60	Male	Calf	Unknown	Metastasis	Fatal
Kinoshita and Yasoshima [14] (2007)						

the time. In contrast to chordoma, the matrix of parachordoma is abolished by hyaluronidase predigestion. Additionally, fluorescence in situ hybridization for the

EWSR1 gene break-apart rearrangement is positive in extraskelatal myxoid chondrosarcoma but not in parachordoma.

The existing English literature reveals 45 cases of parachordomas [1–23]. Two of these previously reported cases have somewhat ambiguous pathology that may be more consistent with chordoma, rather than parachordoma [3, 17]. A review of the studies reveals parachordoma has a slight male predilection, with 25 of 44 (56.8%) documented cases occurring in males and 19 of 44 (43.2%) cases occurring in females (Table 1). The average age of the patients is 34.4 years (range, 4–86 years). Of the case reports that mentioned location, the lower extremity was the most common location, with 21 of 41 (51.2%) cases occurring there. Eleven cases occurred in the upper extremity (26.8%). Eight (19.5%) cases occurred in the thorax, trunk, or pelvis. One case occurred in the nares (2.5%).

Parachordoma is considered a slow-growing tumor of an indolent, less aggressive nature than chordoma. There has been reported late recurrence in nine of 45 cases anywhere from 3 months to 12 years later [7, 14, 15]. However, it is difficult to extrapolate a recurrence rate from these numbers because many patients in the case reports were not followed for a sufficiently long time. Furthermore, most studies do not mention if adequate margins were obtained. In our case, intraoperative frozen section and final pathologic review showed negative margins. The possibility of a late recurrence of disease is something that should be considered when following a patient with parachordoma.

There have been five reported cases of metastases from parachordoma. The first report by Miettinen et al. [17] describes a “chordoma-like” sarcoma in a 67-year-old woman with metastatic disease to the lung who died of the disease 11 months after presentation. However, the pathologic features were more consistent with chordoma than parachordoma. Carstens [3] described a metastatic case arising in the buttocks with widespread disease, with death resulting 14 months later after local recurrence. However, again, the pathology was more like chordoma than parachordoma. These two studies were included previously in the parachordoma literature, but, given the inconsistent nature of the pathology, we believe they should be discounted.

Limon et al. [16] illustrated a case of lymph node metastases from a chest wall parachordoma. The metastatic cells were described as being more cellular and pleomorphic. The fourth metastatic case by Abe et al. [1] reported multiple metastases in a 68-year-old man with a confirmed parachordoma originating in the calf. The patient eventually died 32 months after surgery from disease despite below-knee amputation, radiotherapy, and chemotherapy. The most recent report of metastatic disease is from Kinoshita and Yasoshima [14]. The patient was a 60-year-old man with primary parachordoma in his left calf who died 4 months after surgery from lung and brain metastases.

The case presented here, a 60-year-old woman with a parachordoma of the right humerus, adds to the literature involving this rare tumor. Parachordoma generally is considered a benign tumor with few metastases. However, review of the literature shows there have been at least two confirmed deaths from metastatic parachordoma. Possibly, parachordoma should be thought of as a potentially aggressive low-grade sarcoma. Given this, we performed wide excision of the mass and will continue to follow the patient with routine imaging studies as we would for a patient with a soft tissue sarcoma. To clarify the behavior of this rare tumor, long-term followup studies are needed.

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