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CASE REPORT

Giant Cell Fibroblastoma in a 3-Year-Old Boy

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Giant cell fibroblastoma (GCF) is a rare soft tissue tumor most often discovered during the first two decades of life. We present a case of a 3-year-old boy with a history of a recurrent lesion in the knee, the tumor growth progressively and enlarged to 2.1 cm in the previous two years before diagnosis. It involved the subcutaneous tissue, had infiltrative borders and extended into the superficial dermis. The tumor was surgically excised with free margins. There was no evidence of local recurrence, and a metastatic workup was negative after 10 years of follow up. We review herein the clinicopathologic features, histogenesis, differential diagnosis and relationship to dermatofibrosarcoma protuberans (DFSP). (Pathology Oncology Research Vol 9, No 4, 249–251)

Keywords: tumor, giant cell, giant cell fibroblastoma, dermatofibrosarcoma protuberans

Introduction

Giant cell fibroblastoma (GCF) is an uncommon fibrohistiocytic tumor first described in 1982.[1] It appears as a painless, slowly enlarging, subcutaneous mass and is characterized by the mixed proliferation of fibroblastic cells and multinucleated giant cells within a myxoid or collagenous stroma, including pseudovascular tissue spaces.[2] GCF and DFSP are considered fibrohistiocytic tumors of intermediate grade of malignancy with high incidence of recurrence and with similar biological behavior. Furthermore composite tumors with both components have been reported.[3]

Case report

A 3-year-old boy presented at hospital para el Niño Poblano with a history of a recurrent lesion in his knee. At the age of 1 year-old a 0.4 cm nodule was noted in his extremity. Over the following 2 years the mass progressively enlarged to 2.1 cm. It involved the subcutaneous tissue, had infiltrative borders and extended into the superficial dermis. The patient was asymptomatic and free of any other lesion. The tumor was surgically excised with free margins. There was no evidence of local recurrence, and a metastatic workup was negative after 10 years of follow-up.

Grossly the tumor was grayish-pink with a gelatinous consistency. Microscopically the neoplasm was composed by spindle cells and multinucleated giant cells in a fibromyxoid background *(Figure 1)*. The spindle cells were arranged diffusely or in vague fascicles, their nuclei were elongated and had vesicular to hyperchromatic chromatin. The cytoplasm was scanty and eosinophilic. The giant cells were scattered throughout the tumor, they contained a variable number of round to oval nuclei and had abundant amphophilic cytoplasm with irregular cytoplasmic contours *(Figure 2)*. Pseudovascular spaces were identified and were considered characteristic of this tumor. Mitotic figures were rare, and there was no vascular invasion or tumor necrosis. Immunoperoxidase stains demonstrated that multinucleated giant cells were CD 68 positive, but CD 34 negative. The spindle cell population was positive for CD 34 and negative for S-100, Factor VIII and CD 31.

Ultrastructurally the spindle cells had abundant rough endoplasmic reticulum with variable degrees of dilatation. Cell membranes had numerous irregular cytoplasmic projections and multinucleated giant cells revealed segmentation of the nuclei.

Discussion

GCF is a rare neoplasm occurring almost exclusively in the first two decades of life, about 50% of the cases recur locally and not infrequently is misdiagnosed as a sarcoma

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with the subsequent aggressive treatment.[2] Since the initial description by Shmookler and Enzinger[1] in 1982, less than 100 cases of GCF have been reported. It is believed that examples of this tumor were diagnosed as a low-grade sarcoma in the past.

GCF occurs in the soft tissues, may extend to the subcutaneous region and have a predilection for the thigh, inguinal region, and chest wall. In the 67 cases of GCF reviewed by Maeda et al.[4] in 1998, patient ages ranged from birth to 64 years, with 49 of 67 patients (73.1%) being younger than 10 years of age. Forty four of the 67 cases of GCF occurred in boys and the tumor size ranged from 0.8 to 8 cm. In the case series reported by Shmookler et al.,[2] the preoperative duration of the lesions ranged from 1 to 30 months (average 10.9 months), with 42-50% recurring locally after excision. Although this tumor is locally aggressive, no metastases have yet been reported.

Grossly, GCF is an unencapsulated, gray-white and most often gelatinous mass. Histologically GCF is characterized by diffuse infiltration of the mid-and lower dermis with bland spindle cells and multinucleate giant cell dispersed in a fibromyxoid to hyalinized stroma. There are rare mitotic figures. Irregular pseudovascular spaces lined by a discontinuous row of multinucleated cells are a distinctive feature of GCF. The solid areas contain elongated to stellate cells set within a stroma that varies in cellularity from hypocellular with abundant myxoid stroma to moderately cellular with less myxoid material. Immunohistochemically, the only antigen that has been found to consistently stain GCF has been vimentin.[5] However, the CD 34 antigen can be present not only on spindle-shaped cells but also on some giant cells of GCF.[6] Despite the histogenesis of GCF is currently a subject of debate, fibroblastic differentiation is demonstrated ultrastucturally by encountering multisegmented nuclei, cytoplasmic projections and well developed rough endoplasmic reticulum.[7]

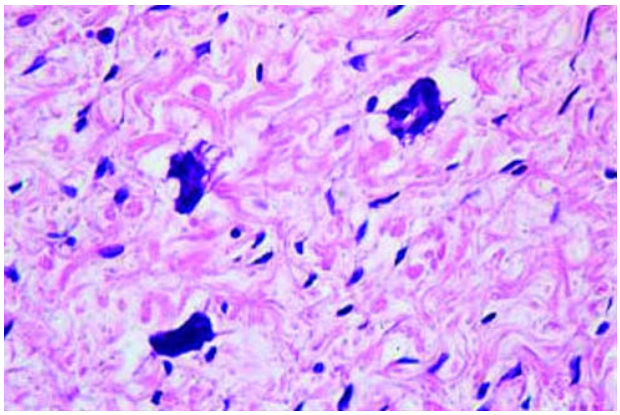

Figure 1. *Spindle and multinucleated giant cells scattered throughout the tumor.*

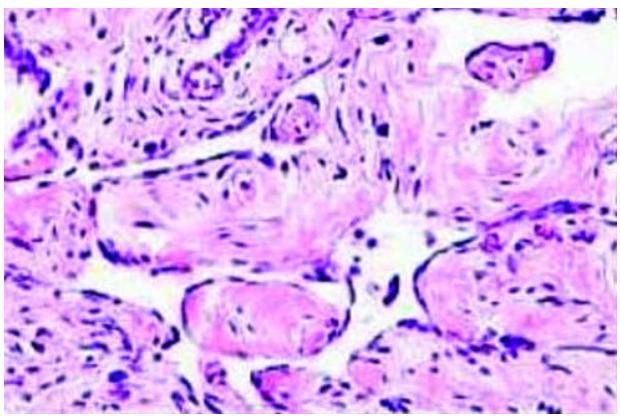

Figure 2. *Pseudovascular tissue spaces lined by multinucleated giant cells.*

The clinical differential diagnosis of GCF includes hypertrophic scar, keloid, dermatofibroma, DFSP, fibrosarcoma, atypical fibroxanthoma and nodular fasciitis.[8] Histologic differential diagnosis should include angiosarcoma, lymphangioma, infantile myofibromatosis, myxoid liposarcoma, and myxoid malignant fibrous histiocytoma. The vascular spaces of lymphangiomas are usually surrounded by lymphocytes and mural smooth muscle is present around the vascular spaces. Giant cells are rare in infantile myofibromatosis and the CD 34 antigen is negative. Angiosarcoma, myxoid liposarcoma and myxoid malignant fibrous histiocytoma are extremely rare in the pediatric group.

GCF and DFSP share several histological features, both lesions are characterized by a proliferation of spindle and stellate-shaped cells within the dermis and there is infiltration of the subcutaneous tissue. The CD 34 antigen that is a very good marker of DFSP is also positive in GCF.[9] In the last years there has been some case reports of GCF associated with DFSP and vice versa, DFSP with areas resembling GCF providing evidence of the close relationship between these two neoplasms.[4,10] Furthermore GCF and DFSP present unique cytogenetic features, such as the reciprocal translocation t(17;22) or more commonly supernumerary ring chromosomes containing sequences from chromosomes 17 and 22.[11] This translocation has been observed also in hybrid lesions suggesting a common biologic process with varying morphologic expression.[12]

In conclusion pathologists should be aware that GCF and DFSP occur in childhood, both lesions are closely related not only histologically but also molecularly and should be placed within the same category of soft tissue tumors.

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