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Aneurysmal ("Angiomatoid") Fibrous Histiocytoma in a Child

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A case of aneurysmal ("angiomatoid") fibrous histiocytoma (AFH) in a 12-year-old girl is presented with its unusual clinicopathologic features. The lesion had the full microscopic characteristics of AFH described in prior reports, but it also had some features that differed from the original description of the disorder, such as the involvement of subcutis, its occurrence in the scalp, and a documented history of minor trauma. The lesion clinically resembled the gross features of hemangioma. The experience in the present case raises the need for considering AFH as one of major differential diagnosis of nodular cutaneous tumors in children that simulate malignancy such as angiomatoid malignant fibrous histiocytoma and Kaposi's sarcoma.

Key Words: *Aneurysmal fibrous histiocytoma, Neoplasm, Skin.*

INTRODUCTION

Fibrous histiocytoma of the skin is a category of benign tumors of fibrohistiocytic origin, showing proliferation of fibroblastic or histiocytoid spindle cells in storiform arrangement. The term "Sclerosing hemangioma" has been used to designate those cutaneous fibrous histiocytomas whose main portions were composed of capillaries (Enzinger and Weiss, 1988).In 1981, Santa Cruz and Kyriakos described a special variant of cutaneous fibrous histiocytoma which was characterized by large, blood-filled cystic spaces and diffuse hemosiderin deposits. They proposed the term "Aneurysmal (Angiomatoid) Fibrous Histiocytoma of the Skin (AFH)" for the subtype as a benign counterpart of angiomatoid malignant fibrous histiocytoma (AMFH) previously described by Enzinger (Enzinger, 1979). Recently, we experienced a case of AFH with typical gross and microscopic features but with unusual clinicopathologic manifestations, and we believe that the unusual findings deserve some mention.

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CASE REPORT

This 12 years old girl sustained a minor trauma to the right parietal region in 1985, and after that event, a bean-sized, non-tender mass developed in the same area. It slowly increased in size, and an excisional biopsy was performed at a local clinic in 1986. The tissue diagnosis of the lesion was not obtained at the time. About a year later, a small mass recurred in the same area and again began to increase in size slowly. So her mother brought her to the Department of Neurosurgery of Seoul National University Children's Hospital for further treatment. On physical examination, a 3cm×3cm-sized protruding mass with overlying purplish discoloration was found in the scalp. The mass was soft and non tender. Other laboratory findings were unremarkable, and there was no associated systemic manifestation. Under the clinical impression of hemangioma, an operation was done. The tumor was easily separated from the underlying periosteum and completely extirpated.

PATHOLOGICAL FINDINGS

The received specimen was a portion of the scalp with a tumor and measured 4cm×3.5cm×1.3cm. The epidermis showed a well circumscribed, slightly pro-

truding purplish nodular mass with a crusted and ulcerated lesion on its surface. A 0.6cm long old scar was present on the periphery of the lesion. The cut surface revealed a well-delineated lesion with characteristic, chocolate-like, gelatinous cut surface and partial cystic change (Fig. 1). The tumor mass measured 2.7cm×1.5cm in maximal dimension and involved both dermis and subcutis, but there was no evidence of infiltrative foci into the surrounding tissue.

On microscopic examination, the mass was composed of fairly cellular portions and large, blood-filled cystic spaces of variable size. The individual cells had bland looking ovoid nuclei and elongated cytoplasm.

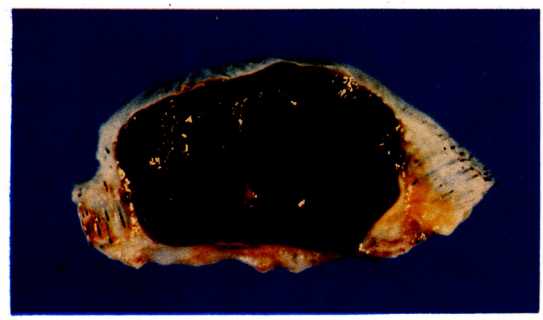

Fig. 1. The characteristic, chocolate-like gelatinous cut surface of the mass. It involves lower dermis and is relatively well circumscribed from surrounding soft tissue.

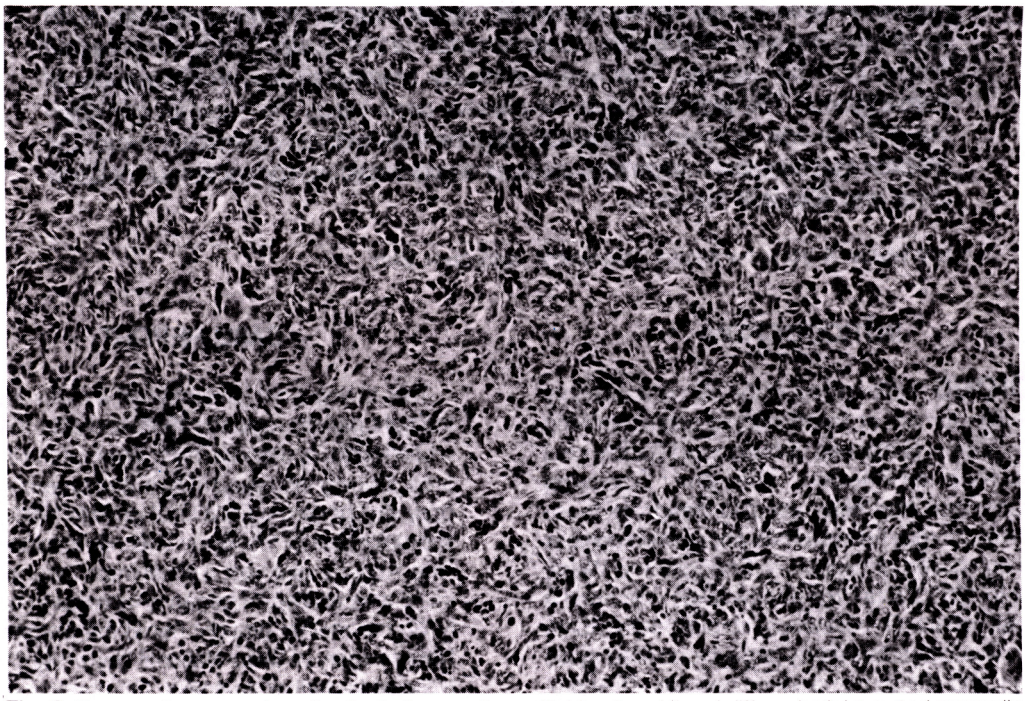

Fig. 2. Tumor cells arrange in a classic storiform pattern with bland nuclei and diffuse, both intra- and extracellular hemosiderin deposit.

The arrangement of the tumor cells was mainly of classic storiform pattern (Fig. 2). Multinucleated Touton-type giant cells were also frequently found. The blood-filled clefts or cystic spaces were mostly devoid of definite endothelial linings while having features of blood-filled cracks (Fig. 3). Another characteristic feature of the lesion was diffuse, intra- or extra-cellular deposition of hemosiderin pigment that showed strong positive reaction to iron staining (Fig. 4). The mitotic figures were generally negligible, but areas with a mitotic count up to 3/10HPF were present in the periphery of the tumor. A focal, dense dermal fibrosis suggested a change related to the previous biopsy. The mass occupied, as found on gross examination, most of the dermis and subcutis except for the upper portion of the dermis. The epidermis showed mild acanthosis. Immunohistochemical staining to S-100 protein and lysozyme, using the avidin-biotin complex method, gave complete negative reaction to the tumor cells.

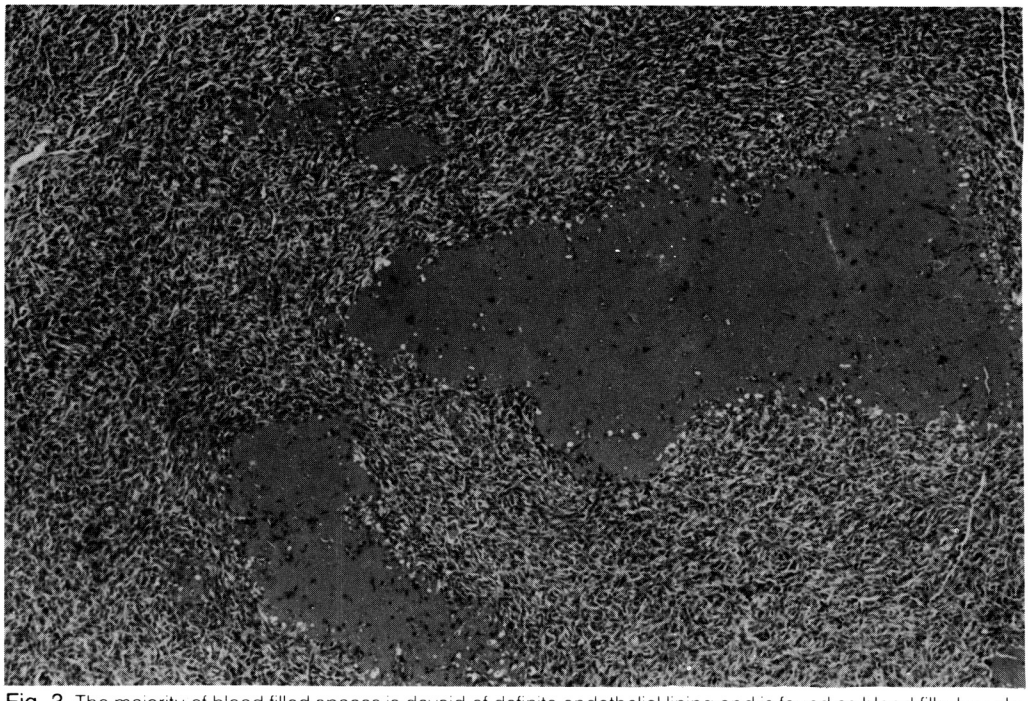

Fig. 3. The majority of blood-filled spaces is devoid of definite endothelial lining and is found as blood-filled cracks or clefts.

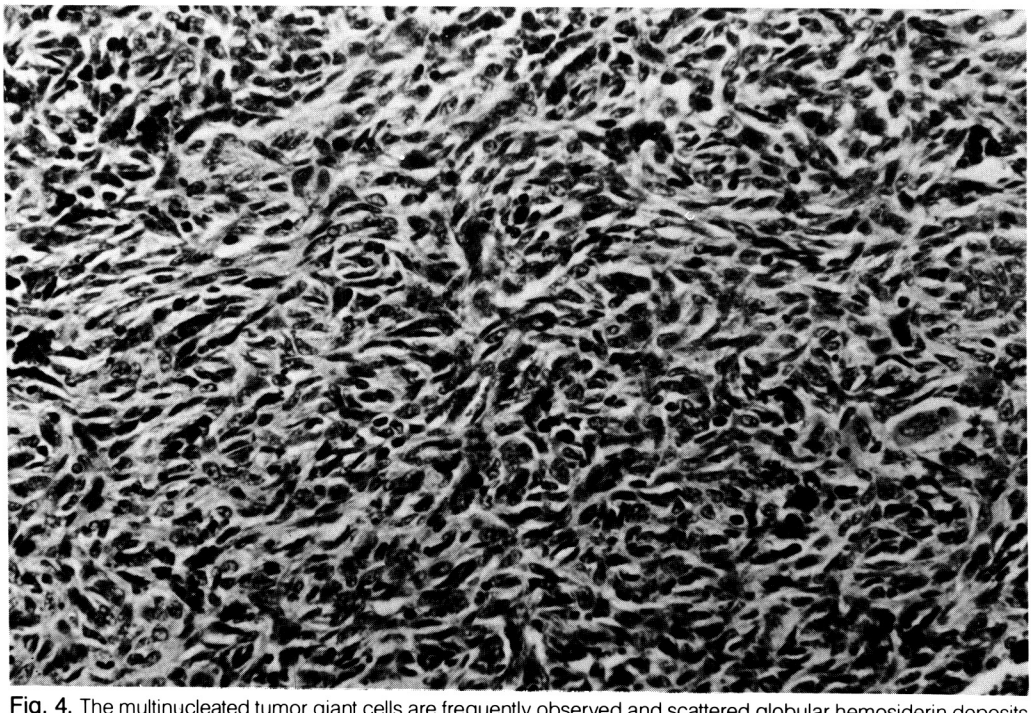

Fig. 4. The multinucleated tumor giant cells are frequently observed and scattered globular hemosiderin deposits are characteristic.

DISCUSSION

Kauffman and Stout (1961) reviewed their experience in histiocytic tumors in children and broadly categorized them into fibrous xanthomas and fibrous histiocytomas. They thought that histiocyte was the basic origin of these tumors and regarded histiocyte as one of the mesenchymal cells that is capable of acting as a facultative fibroblast. They pointed out that the great majority of these tumors is benign and did not exhibit any evidence of biological malignancy.

In 1979, Enzinger described 41 examples of an unusual sarcoma of fibrohistiocytic origin, which were characterized by their primary occurrence as a nodular subcutaneous growth in the extremities or, more rarely, in the head and neck of young individuals and children. He named those cases "Angiomatoid Malignant Fibrous Histiocytoma" (AMFH). Their distinctive microscopic features included solid arrays or nests of fibroblast- and histiocyte-like cells, containing varying amount of intracellular hemosiderin or lipid, focal areas of hemorrhage or hemorrhagic cyst-like spaces, along with aggregates of chronic inflammatory cells. Histories of trauma were definite in a significant portion of cases. About two years later, Santa Cruz and Kyriakos (1981) reported 17 cases of a variant of cutaneous fibrous histiocytoma having different features from ordinary fibrous histiocytomas under the name of "Aneurysmal (Angiomatoid) Fibrous Histiocytoma of the Skin" (AFH). The cases had microscopic resemblance to AMFH, and they considered AFH to be a benign counterpart of AMFH. AFH differed from AMFH in its absence of definite trauma history, its main occurrence in middle-aged population, its lack of inflammatory infiltrate, no cellular pleomorphism, and lack of associated systemic manifestations such as anemia, chill, pyrexia, etc. The mitotic count didn't significantly contribute to the differential diagnosis of AFH and AMFH.

The present case had rather typical histologic findings of AFH but shared some clinicopathologic resemblance to AMFH according to previous descriptions, such as its occurrence in a child (the actual onset ot the disease probably occurred when the patient was only 9 years old, although the tissue diagnosis was not obtained at the time), its location in the scalp, the involvement of the subcutis, and the documented history of trauma.

Besides AMFH, the major differential diagnosis of this case included Kaposi's sarcoma and Bednar tumor (Enzinger and Weiss, 1988). The nature of the pigment is a critical one since the Bednar tumor contains melanin. The presence of acanthotic epidermal change and foam cells in the tumor minimized the possibility of the Kaposi's sarcoma, and slit-like vascular lumina or cellular atypism present in Kaposi's sarcoma were not found. Sood and Mehregan (1985) reported factor VIII antigen as a helpful marker in differentiating AFH from Kaposi's sarcoma.

In summary, though we agree with the view of Santa Cruz and Kyriakos (1981) that AFH lies in the spectrum of entities such as sclerosing hemangioma and hemosiderin histiocytoma (Bernstein, 1939) and that the intratumoral hemorrhages and hemosiderin deposits are related to repeated minor trauma, we believe that the aforementioned different features of AFH in this case should be added to those in the original description. AFH should be considered as one of the differential diagnosis of cutaneous nodular tumors in children, and its particular importance lies in that it may be misdiagnosed as aggressive conditions, including AMFH or Kaposi's sarcoma, with which surgical pathologists are more familiar.

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