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Aneurysmal Fibrous Histiocytoma: Clinicopathology Analysis of 30 Cases of a Rare Variant of Cutaneous Fibrohistiocytoma*

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Summary: Aneurysmal fibrous histiocytoma is often clinically misdiagnosed. In this study, we put forward an insight on how to help diagnose this disease clinically. A retrospective chart review was performed on all patients diagnosed with aneurysmal fibrous histiocytoma from 2007 to 2017 in the Department of Dermatology, Union Hospital, China, and all clinical data were collected from the hospital archives. From a total of 418 patients diagnosed with cutaneous fibrous histiocytoma, only 30 patients were confirmed to have aneurysmal fibrous histiocytoma out of which only 2 patients were clinically diagnosed with aneurysmal fibrous histiocytoma. The remaining 28 patients were diagnosed with various types of vascular tumors although pathology classified them as having aneurysmal fibrous histiocytoma. Among the 30 patients, 9 were male and 21 were female. There were following age groups: 13–19 (mean 16, *n*=4), 20–29 (mean 26.25, *n*=8), 30–39 (mean 33, *n*=7), 40–49 (mean 44, *n*=4), 50–59 (mean 56.75, *n*=4), 60 and above (mean 61, *n*=3). Tumors were present on the head, neck, back, waist, hips and upper and lower extremities. After complete excision, there was no recurrence and no complications. Histologically, lesions showed the typical pseudoangiomatoid spaces without endothelial lining and infiltration of fibrohistiocytes in hemosiderotic pigmentation. It was suggested that although the prognosis of aneurysmal fibrous histiocytoma is good, accurate diagnosis is paramount to avoid clinical misdiagnosis and subsequent complications.
Key words: aneurysmal fibrous histiocytoma; cutaneous fibrous histiocytoma

Aneurysmal fibrous histiocytoma is a rare variant of cutaneous fibrous histiocytoma. It's a benign tumor mostly in young/middle-aged adults with a higher incidence in females than males[1]. The lesions typically present with various colors, morphology, and texture. It may manifest as blue-black, yellow, red or skin colored. Among the differential diagnoses that may mimic aneurysmal fibrous histiocytoma on presentation, there are Kaposi sarcoma, malignant melanoma, hemangioma, spindle cell hemangioma, neurofibroma, angiosarcoma, and aneurysmal malignant fibrous histiocytoma. Histologically, aneurysmal fibrous histiocytoma may be confused with dermatofibrosarcoma protuberans, non-specific cysts[2].

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The typical histologic presentation of aneurysmal fibrous histiocytoma is whorls of the spindle and stellate cells in an un-organized pattern, irregular cavities containing red blood cells without endothelial lining, fibrous pseudo capsule[3]. Here we presented a total of 30 cases of aneurysmal fibrous histiocytoma with a focus on the clinical and histopathological analysis of the features of this rare skin tumor and some important differential diagnosis.

1 MATERIALS AND METHODS

We reviewed patients' data retrospectively who'd initially been diagnosed with the following medical conditions: cutaneous fibrous histiocytoma, fibroma, aneurysmal fibrous histiocytoma, dermatofibroma and angiomatoid fibrous histiocytoma based on the data extracted from the database of Department of Dermatology, Union Hospital, China between 2007

and 2017. All clinical data and pathology reports were extracted from the hospital archives.

2 RESULTS

From a total of 418 patients diagnosed with cutaneous fibrous histiocytoma, only 30 patients (i.e., both clinically and pathologically) were confirmed to have aneurysmal fibrous histiocytoma, out of which only 2 patients were clinically diagnosed with aneurysmal fibrous histiocytoma. The remaining 28 patients were diagnosed with various types of vascular tumors [nevus (n=4), dermatofibroma (n=13), plexiform neurofibroma (isolated) (n=1), mesenchymal tumors (n=4), targetoid hemosiderotic hemangioma (n=1), epidermal cyst (n=2), fibroxanthoma (n=1), hemangioma (n=1) and tumor (n=1)]. But the pathology had classified them as being aneurysmal fibrous histiocytoma. Among the 30 patients, 9 were male and 21 were female with ages ranging from 13–63 (median 31.5 years) (table 1).

The tumors in all the 30 patients were predisposed on the head, neck, back, waist, hips, and upper and lower extremities (fig. 1A–1D). All tumors were completely excised with clear margins (an approximate of 0.5 mm away from cancer). After complete excision, recurrence and complications were not reported. Pathologically, pseudospaces without endothelial lining filled with blood together with multiple clefts were observed. Hemosiderin deposition embedding the proliferated fibroblasts and histiocytes was seen in the dermis which is not a typical presentation of cutaneous fibrous histiocytoma, hence confirming aneurysmal fibrous histiocytoma.

Microscopically, we observed hyperplasia, acanthosis, lowering of rete ridges, and pigmentation of the basal layer in the epidermis while in the dermis, we saw the proliferation of collagen fiber and fibroblasts in palisading arrangement forming large clefts with no endothelial lining but filled with erythrocytes. Moreover, no nuclear atypia was observed in all cells (fig. 1E–1I).

3 DISCUSSION

Aneurysmal fibrous histiocytoma is a fibro-histiocytic tumor classified into the architectural peculiarities category[4]. Lesions have a variety of

Table 1 A summary of the clinical data of the 30 patients with aneurysmal fibrous histiocytoma confirmed by pathology

Sex/Age	Presentation and size/cm	Initial diagnosis
F/36	Hard black papule at the back of the neck, 4 yrs/0.8	Atrophic dermatofibroma
F/19	Yellow nodule on the left upper arm, 2 yrs/0.6	Dermatofibroma
F/59	Painful flesh colored cyst on the right medial wrist, 6 yrs/1.5	Schwannoma, vascular leiomyoma
F/63	Yellowish papule on the outer left thigh, 1 yr/1	Dermatofibroma
F/22	Black papule on the right lower leg, 3 months/0.8	Intradermal nevus
M/31	Flesh colored nodule on the right upper arm, 1 yr/1	Dermatofibroma
F/41	Papule on the right hip, previously treated with laser, 2 months/0.5	Melanocytic nevus
F/59	Red, pink nodule on the right hand, 2 months/0.5	Dermatofibroma
M/47	Pink nodule on the right upper arm, 5 yrs/1.2	Dermatofibroma
M/31	Pea-sized nodule on the right upper gluteal area, 8 yrs/1.4	Intradermal nevus
F/30	Painful papule on the lower leg, 3 yrs/1.2	Leiomyoma
M/48	Flesh colored nodule on the right lower leg, 3 yrs/1.4	Dermatofibroma
F/29	Brown nodule at the back, 1 yr/0.6	Dermatofibroma
F/13	Pink papule at the lower thigh, 4 yrs/1	Dermatofibroma
F/60	Black papule on the inner left thigh with purple border, 2 yrs/1	Targetoid hemosiderotic hemangioma
F/26	Pea-sized nodule on the inner left thigh, 1 month/0.8	Dermatofibroma
F/28	Black nodule on the left upper arm, 1 yr/1.8	Tumor
M/22	Brown painful papule on the waist, 8 yrs/1.4	Dermatofibroma
F/32	Itching nodule on the lower leg, 2 yrs/1.6	Plexiform neurofibroma
M/19	Black papule on the left upper arm, 4 yrs/1	Dermatofibroma
F/39	Painful nodule on the left lateral thigh, 10 yrs/1.2	Epidermal cyst
M/28	Flesh colored papule on the left upper arm, 1 yr/0.6	Dermatofibroma
M/59	Pink papule on the right knee, 3 yrs/1.6	Fibroxanthoma
F/60	Painful red cyst on the waist, 2 yrs/2.2	Rhabdomyosarcoma
F/50	Tender papule on the lower left leg, 1 yr/1	Dermatofibroma
F/32*	Nodule on the left upper knee, 2 yrs/1	Aneurysmal fibrous histiocytoma
F/27	Painful violaceous papule on the left upper arm, 1 yr/0.8	Nevus with secondary infection
F/40*	Yellowish nodule on the left thigh, 10 yrs/2	Aneurysmal fibrous histiocytoma
M/13	Hard black, brown papule on the occipital area, 1 yr/0.8	Epidermal cyst, pigmented nevus
F/28	Red papule on right upper arm, 10+ yrs/2	Hemangioma

*Accurate diagnosis before pathology. The table includes all diagnoses and, in some patients, more than one diagnosis was made.

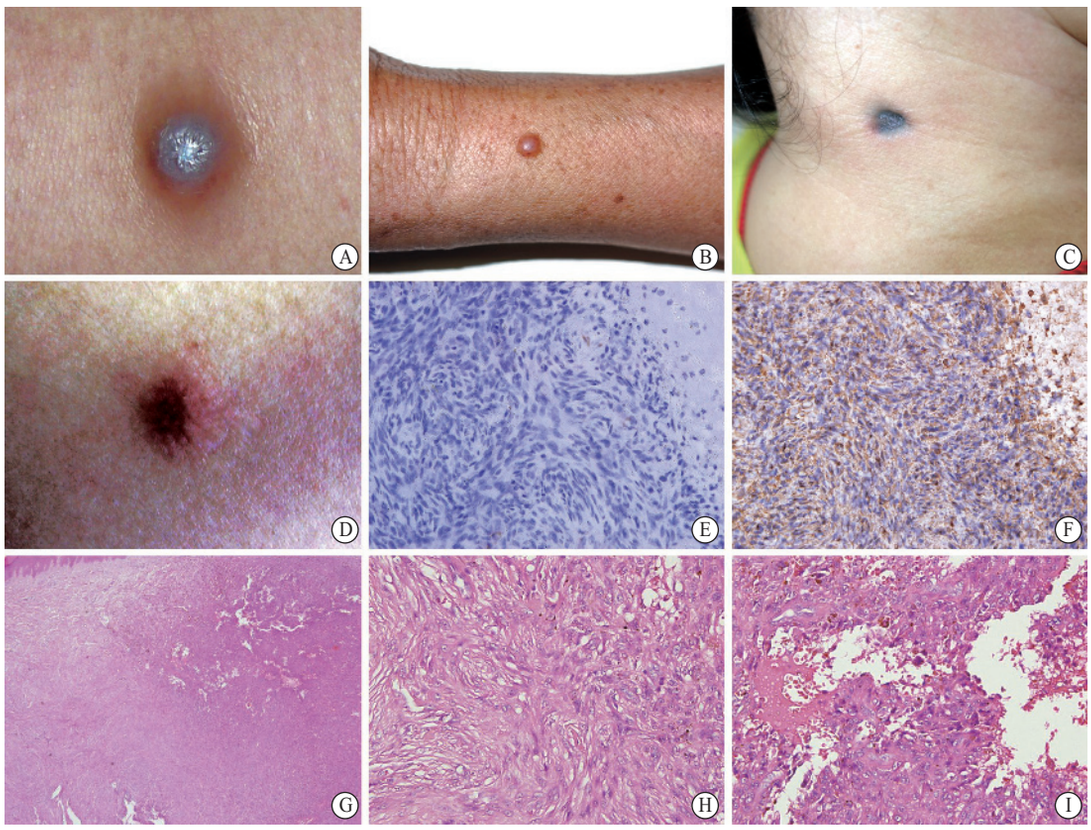

Fig. 1 A–D: various easliy misdiagnosed lesions with different sizes and colors. E and F: Immunohistochemical stain (demonstrated by actin) of fibroblasts was negative (E, ×400) and vimentin positive (F, HE ×400). G: The tumor was mostly in the dermis consisting of spindle cells with clefts (HE ×40). H: storiform pattern (whorly arrangement) of fibroblasts and histiocytes (HE ×400). I: irregular cavities without endothelial lining with extravasated erythrocytes and hemosiderin deposition (HE ×400)

clinical presentations and may mimic other benign and malignant tumors[5], thus making it harder to diagnose clinically.

The lesion appears as a polypoid, smooth, slow growing, larger than cutaneous fibrous histiocytoma and mostly located on extremities. It may present with different colors with multinodular or cystic texture, with or without pain due to abrupt hemorrhage[6] and its indolent nature makes it less aggressive than angiomatoid fibrous histiocytoma.

The differences in clinical presentation of aneurysmal fibrous histiocytoma from benign cutaneous fibrous histiocytoma are due to the slow continuous extravasation of blood in the tumor[7] which gives it a blue-black appearance. The lesions may turn yellow according to the ratio of lipophages to siderophages in the infiltration. They achieve flat morphologies due to fibrosis of the papillary dermis and myofibroblast contraction with decreasing cellularity[8], thus making it a challenge to diagnose the disease clinically.

Furthermore, we suggest that the differences in the color presentation may depend on the severity of focal thrombosis and quantity of hemosiderin in the cytoplasm.

It should also be noted that aneurysmal changes are sometimes seen in other benign subtypes of fibrous histiocytoma such as the cellular and atypical variants.

Our study found that all tumors were completely excised with clear margins (approximately 0.5 mm away from a tumor). We noted no recurrence and metastasis, which is inconsistent with that described by Shi *et al* who reported higher recurrence and metastasis in their patients[9].

Histologically, aneurysmal fibrous histiocytoma shows intralesional pseudo cysts filled with blood and no endothelial lining surrounded by spindle cells arranged in a storiform pattern. There's proliferation of fibroblasts and histiocytes in capillary rich stroma[10], foamy macrophages, abundant hemosiderin and dilated blood vessels. Cytologically, it shows multi-nucleated cells, a cellular smear of spindle cells with hemosiderin pigmenation[5, 7]. Atypia of the nucleus is not definite in both histology and cytology. It has been shown that aneurysmal fibrous histiocytoma is usually negative for CD34, CD31, CD68, desmin and factor Ⅷa in the stromal cells[2] but positive for vimentin[9, 11]. The importance of biopsy is to differentiate aneurysmal fibrous histiocytoma from its differentials.

However, the difference between pigmented dermatofibrosarcoma protuberans and aneurysmal

fibrous histiocytoma is, the latter occurs in subcutis, shows no reticulation in the fatty layer and stains positive with CD34[11].

Spindle cell melanoma is differentiated from aneurysmal fibrous histiocytoma by performing a cellular smear which shows multi-nuclei, macronucleoli and intranuclear pseudoinclusions.

Kaposi sarcoma can be differentiated from aneurysmal fibrous histiocytoma since it is HHV8 positive, and found on extremities presenting as violaceous papules in older individuals. Microscopically, in Kaposi sarcoma, there is the endothelial lining of the enlarged clefts rather than blood spaces and stains positive for CD34 and Ⅷ[8, 11–13]. Spindle cell hemangioma is located in the subcutis, poorly circumscribed and consists of true vascular capsules lined with endothelium[14].

Angiomatoid fibrous histiocytoma differs from aneurysmal fibrous histiocytoma because of the presence of myxoid structure and fibrous pseudo capsule and occurs mostly on extremities in the young[15, 16].

Angiosarcoma differs from aneurysmal fibrous histiocytoma since it occurs on the face and scalp of elderly and in those with a history of radiation therapy. Furthermore, angiosarcoma is known to occur in a subcutaneous layer with the atypical proliferation of endothelia[4, 9, 11, 14].

In conclusion, although the prognosis of aneurysmal fibrous histiocytoma is good, careful and accurate diagnosis is required to avoid the misdiagnosis of malignant vascular tumors. It's also advisable to have a clear grasp on the different variants of cutaneous fibrous histiocytoma and their classifications since they have different rates of recurrence, similar biological constitution and histologically present with typical components of the lesion.

Conflict of Interest Statement

The authors declare that they have no competing interests.

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