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Neck Dermatofibrosarcoma Protuberans: A Rare Children Cutaneous Tumor

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Abstract:
Dermatofibrosarcoma protuberans (DFSP) is an uncommon, locally aggressive, cutaneous tumor which rarely metastasizes. However, local recurrence following apparently adequate surgical excision is well recognized. Its occurrence in children head and neck region is very rare. We present a case of children DFSP with oncological, functional and aesthetic challenges that arose from it. The patient was treated with a wide excision of the lesion. One year later, there was no recurrence. We also reviewed the relevant literature on DFSP pertaining the head and neck region and discussed the diagnosis and treatment based on that literature.

Keywords: *dermatofibrosarcoma protuberans, diagnosis, treatment, local recurrence, children*

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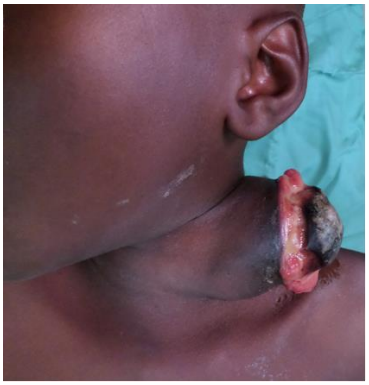
I. Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon, locally aggressive, soft tissues malignant tumor characterized by high propensity for local relapse and low metastatic potential. Head and neck cases accounted for approximately 10 -15% of cases.[1-4], although DFSP represented 1% of all soft tissues sarcomas and less than 1% of all head and neck malignancies.[5] It's reported to occur rarely to children.[6] Our case highlights two specific features. First, understanding the natural history of this entity is crucial even though there were potential difficulties in the diagnosis of DFSP owing to its rarity. Secund, it's functional and aesthetic reconstruction with the risk of local recurrence.

II. Case report

An eleven years old child presented a mass infiltrating his left lateral neck, which had been slowly growing during five years. Since six months, that mass had quickly increased in size with onset of ulceration and skin necrosis two week before consultation and after applications of plant based mixtures on the mass. The examination revealed a painless mass approximately 6, 3 × 5 cm size, which was hard and not fixed in the left cervical region (figure 1a). No cervical lymph node was found by the neck CT scan. A mass of 6, 7 ×5, 6 × 7, 6 cm size, which was regularly demarcated and had no invasion of surrounding muscles and bone structures, was determined (figure 1b). The rapid evolution of the neck mass and the very limited nature of the latter, the indication of excision biopsy was posed (figure 2a). During the operation, it was observed that the mass involved the cutaneous and the subcutaneous tissues, but not the muscles. By leaving a surgical margin (2, 1 cm), a wide excision was performed on the mass. The skin defect formed after surgery was closed primarily without skin grafting (figure 2b). The postoperative histopathology result was reported as dermatofibrosarcoma of Darier and Ferrand (figures 3). One year after surgery, there was no clinical or radiologic recurrence (figure 5).



Figures 1: (a) clinical image showing a neck lesion (6 × 4 cm in size)

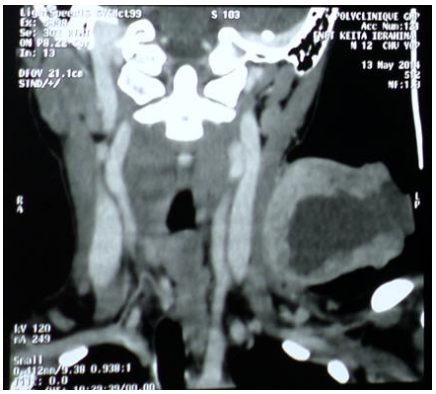


Figure 1.b: Computed tomography image showing a cervical mass adherent to

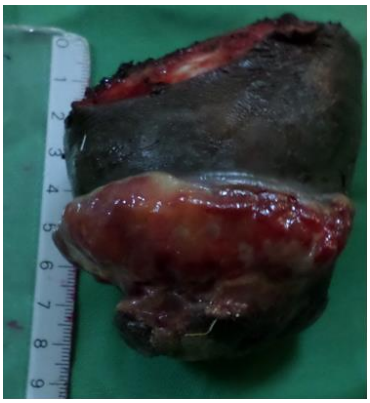


Figure 2.a : Gross specimen. Outer surface of ill-defined ulcerative and necrotizing mass

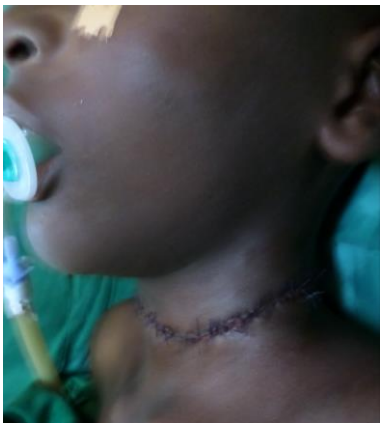


Figure 2.b: postoperative image following local excision and reconstruction

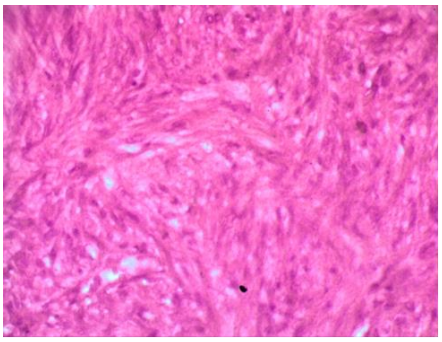


Figure 3: Histopathology image

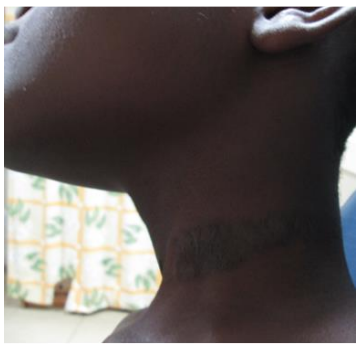


Figure 4: clinical image showing none lesion one year

III. Discussion

Dermatofibrosarcoma protuberans (DFSP) is a rare locally aggressive cutaneous tumor of intermediate malignancy. Although metastasis rarely occurs, DFSP has a locally aggressive behaviour with a high recurrence rate.[7] Head and neck involvement is unusual.[1,7,8] The occurrence of this tumor is most prevalent in the age group 20-40.[1,3] This tumor is rare in childhood and the congenital form is exceptional.[3,9] No evidence of hereditary or familial predisposition exists.[1] A slight male predominance is suggested by other authors and its incidence in black patients is almost twice than the Caucasians.[1,10]

Clinically, the tumor's appearance depends on the stage of the disease. Initially it presents as a cutaneous pink to red-bluish painless plaque which grows more nodular with time and over time into ulcerative hemorrhagic protuberant tumor. It develops superficially, mobile upon palpation as it adhered with its overlying

skin, but not with the underlying tissues. Unfortunately, fixation to deeper structures such as fascia and muscles may occur in the later stage of the tumor. Slow tumor growth is characteristic and there is often a delay in diagnosis as patients can ignore the lesion from weeks to years due to its indolent nature.[1,3] HAMID and Al[11] relieved that the duration of lesions at the time of first presentation was under two years in 68, 88% cases (minimum duration 1 month and maximum 7 years). The decline in the illness duration is probably due to freely available medical facility, increased literacy, and increased in the familiarity with the disease. The precocity of diagnosis is depending upon the location or the size of tumor, level of literacy, and availability of health care facilities. Trauma has frequently been incriminated as a possible factor influencing the growth of the tumor. The relationship between trauma and tumor seems to be only coincidental[11]. In our case, the patient seeks medical attention after five years when the tumor raised and ulcerated. This ambiguous and seemingly benign clinical presentation necessitates making a final diagnosis based on extensive histological analysis. Its differential diagnosis from dermatofibroma is difficult, both clinically and histopathologically. There are a positive reaction with Factor XIIIa antibody and a negative reaction with CD34 antibody.[1,4] Since superficial changes such as erosion and ulceration are rarely seen in dermatofibromas, there is a small amount of literature data on this subject.[12, 13] It is difficult to make differential diagnosis between dermatofibrosarcoma protuberans and ulcerative dermatofibroma, since the size of the lesion is bigger than the classical dermatofibroma, with the localization being in locations other than the lower extremities; furthermore, the histopathological presence of superficial destruction, the intense cellular appearance, the swirl-like distribution and the observation of penetration to the deep dermis and subcutaneous tissue make the differential diagnosis difficult.[13] Immuno-histochemical studies performed with the use of Factor XIIIa and CD34 antibodies may be helpful in such cases.[12,13] A positive staining of CD34 and a negative staining of Factor XIIIa are observed in dermatofibrosarcoma protuberans; a negative staining of CD34 and a positive staining of Factor XIIIa are observed in dermatofibroma.[1,13] The differential diagnosis should also include malignant fibrous histiocytoma, atypical fibroxanthoma, diffuse neurofibroma, giant cell fibroblastoma, myxoid liposarcoma, myxofibrosarcoma and desmoplastic melanoma.[13] Recently, UGUREL and Al[12] have proposed three stages: the stage I represents the primary tumor stadium, the stage II describes a DFSP with regional lymph node metastases and stage III characterizes distance metastases.

DFSP tumors are characterized by unpredictable and widespread subclinical extension, hence high rates of recurrence. The goal remains complete excision of tumor cells with maximum preservation of normal tissues to maintain function and an acceptable aesthetic appearance. Standard treatment of DFSP is a wide local excision with margins between 2 and 3, 5 cm to achieve negative margins for adequate local control.[2,11] We reason that tumors of the head and neck have a higher recurrence rate in the literature because of the tendency of surgeons to take narrower margins at these sites. Difficulties in obtaining wide margins are due to anatomic constraints as well as functional and aesthetic concerns.[1,6,11] Some authors consider the Moh's Micrographical Surgery (MMS) as the treatment of choice in particularly anatomically challenging areas such as the head or neck[11] and in the treatment of children.[1,8, 14] Micrographic surgery uses the microscope to trace out the microscopic ramifications and a map to guide excision of residual tumor. The favorable success rate of MMS in patients with DFSP is attributed to the meticulous procedure of microscopically checking 100% of the tumor margin.[15] MMS and immunohistochemical are not practiced in Côte d'Ivoire. In our case, the treatment of the tumor is a wide and deep local excision (WLE), including the underlying fascia and superficial muscle layer. Since primary closure is not always feasible, reconstructive surgery, using local skin flap, skin grafting, mesh or myocutaneous flap may be required. Neck dissection is not necessary unless a suspicious regional lymphadenopathy is present. Only in DFSP-fibrosarcomatous (FS) cases sentinel lymphnode biopsy is recommended.[1] Although surgery is the gold standard in its treatment, adjuvant radiotherapy has been suggested in unresectable lesions or recurring lesions, when the margins are positive or close to the tumor or in cases where wider excision is not possible due to anatomic limitations, especially in head and neck regions.[16] Chemotherapy with selective tyrosine kinase inhibitory such as Imatinib Mesylate was registred for therapy of inoperable and/or metastatic DFSP. It has also been reported to induce complete or partial remissions in patients with advanced or recurrent DFSP.[17]

IV. Conclusion

DFSP is rare children's head and neck tumor with a high propensity to recur. It poses diagnosis and reconstructive challenges. Based on the best available evidence we recommend a staged excision and reconstruction with at least 2 cm surgical margins. Metastases and multiples recurrences are rare. Clinical and radiological surveillances have to be imposed.

Consent: written informed consent was obtained from the patient for publication of the case reported in this review and accompanying images.

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