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Darier and Ferrand's Dermatofibrosarcoma with Craniofacial Location: A Rare Entity

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Abstract **Case Report**

Introdcuction: Darier and Ferrand's dermatofibrosarcoma is a rare but not exceptional malignant mesenchymentous skin tumor. It represents 0.1% of malignant skin tumors. First described by Jean Darier and Marcel Ferrant in 1924, it is a slow-growing tumor, currently considered to be a true low-grade malignancy fibrosarcoma. The authors report in this work the clinical and evolutionary aspects of three cases of Darier and Ferrand dermatofibrosarcoma located at the cephalic end. ***Observations:*** These were 3 cases of Darier and Ferrand dermatofibrosarcoma encountered between 2010 and 2016. The subjects were female. The ages ranged from 30 to 44 years old. The diagnostic delay varied between 6 and 10 years. The general condition was altered in two cases. The localization was facial in two cases and the scalp in one case. The size of the tumors varied between 15 and 20 cm. All patients underwent maxillofacial computed tomography (CT) and laboratory workup. The clinically suggested diagnosis was confirmed by histological examination. One patient underwent surgical excision with a 3 cm margin. Covering the loss of substance appealed to thin skin grafting. The evolution was favorable after a 7-year follow-up. In the other two cases, the evolution was marked by death before surgical management. ***Conclusion:*** DFS is a rare malignant skin tumor with slow local progression that can often be life-threatening. Early diagnosis ensures efficient treatment.

Keywords: dermatofibrosarcoma, Darier, Ferrand, craniofacial location, rare entity.

INTRODUCTION

Darier and Ferrand's dermatofibrosarcoma (DFS) is a rare but not exceptional malignant mesenchymentous skin tumor, representing 0.1% of malignant skin tumors. The worldwide incidence of DFS is estimated to be 0.8 to 4.2 cases per million population per year. First described by Jean Darier and Marcel Ferrant in 1924, it is a slowly developing tumor, currently considered to be a true fibrosarcoma with low grade of malignancy [1, 2, 3]. This condition occurs quite frequently in men in the 3rd and 4th decades and clinically presents as firm reddish plaques or nodules. The preferred sites are the trunk, extremities, head and neck. It has a good prognosis after complete resection, with a very high risk of local recurrence but with low metastatic potential [3, 4].

The authors report in this work the clinical and evolutionary aspects of three cases of Darier and Ferrand dermatofibrosarcoma located at the cephalic end.

OBSERVATIONS

Clinical case 1

It was about a 30-year-old woman, housewife, married, with no particular medical-surgical history, who consulted for a swelling of the left hemiface in March 2015. The onset of the disease dates back to eight years, marked by the recurrence of a painless swelling of the left hemiface gradually increasing in size. The swelling presented with progressive budding, necrosis and bleeding on contact (Figure 2, 3). The accompanying signs were severe headache and dizziness. On general examination, there was a deterioration in general condition with weight loss and paleness of the skin and mucous membranes. On physical examination, there was a polylobed left hemiface mass with necrosis. They were presented in the form of indurated dermo-hypodermic cupboards,

measuring 195x113x87mm respectively. The lesion was adhering to the left hemiface; it was mobile in relation to the deep plane, painless on palpation. The review of the other devices was unremarkable. The Darier-Ferrand DFS hypothesis was posed. The CT revealed a voluminous process of expansion of the soft parts of the left facial mass without cerebral anomaly. The pre-operative assessment revealed anemia. The biopsy carried out concluded with a DFS of Darier-Ferrand. The patient received a blood transfusion and re-nutrition. The evolution was marked by the death of the patient before surgery.

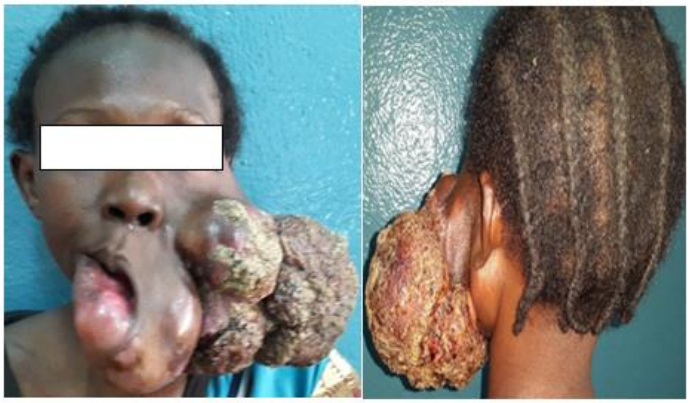

Fig-1 and 2: appearance of the tumor on admission

Clinical case 2

This was a 44-year-old housewife, married patient who presented with a recurrent right hemiface swelling in May 2014 that had progressed for 6 months. She had been operated on 3 times in the past for a left craniofacial swelling, the histological nature of which was not found and had also undergone chemotherapy. The accompanying signs were severe headache, hyperthermia, and dizziness. On general examination, there was a deterioration in general condition with weight loss and paleness of the skin and mucous membranes. On physical examination, there was a swelling of the right hemiface, polylobed, with necrosis. The swelling was gradually budding and bleeding on contact (Figure 3, 4). The lesion adhered to the right hemiface; it was mobile in relation to the deep plane, painless on palpation. The review of the other devices was unremarkable. The Darier-Ferrand DFS hypothesis was posed.

CT revealed a large, heterogeneous polylobed tissue mass implanted in the soft part of the right face, invading the deep planes of the zygomatic fossa and masseter muscles and erosion of the zygomatic arch without cerebral abnormality. The biological assessment objectified a hypochromic microcytic anemia. The patient received a blood transfusion and re-nutrition. The evolution was marked by the death of the patient before surgery.

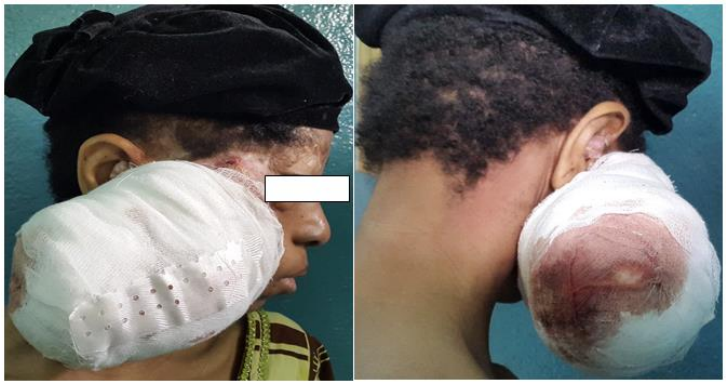

Fig-3 and 4: Appearance of the tumor on admission

Clinical case 3

It was a 35-year-old female patient, housewife, married, fourth gesture, domiciled in Kayes (Mali) with no known particular history who consulted for treatment of a scalp mass in February 2010. The onset of the disease would go back six years, marked by the appearance of a painless right mid-parietal swelling gradually increasing in size. A second ipsi-lateral swelling appeared at the occipital level after six months. Both swellings showed progressive budding (Figure 5). The accompanying signs were severe headache and dizziness. There was no weight loss or fever.

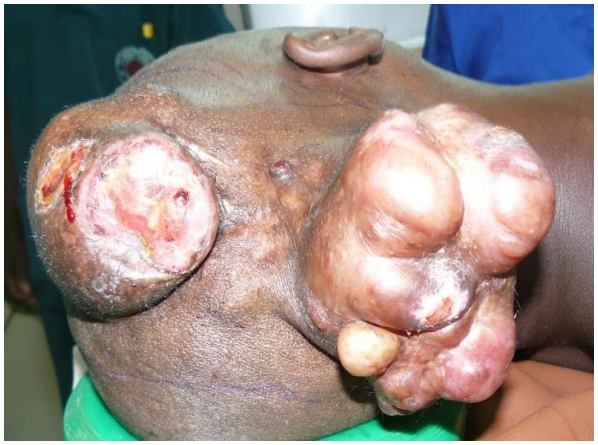

Fig-5: appearance of the tumor on admission

Faced with this picture, the patient had recourse to a traditional treatment which led to ulcerations in places on the two swellings. General signs including blood pressure, pulse, conjunctiva and temperature were unremarkable. On physical examination we found two polylobed right midparietal and occipital masses with ulcers. They were presented in the form of indurated dermo-hypodermic patches measuring 38.3x33 mm and 89.6 x 52.3 mm respectively. These lesions adhered to the scalp, mobile in relation to the deep plane, painless on palpation and associated with local hypoaesthesia. The review of the other devices was unremarkable. We then posed the hypothesis of Darier-Ferrand dermatoneurofibroma. The brain CT scan without and with injection made it possible to objectify an isodense subcutaneous mid-parietal and right occipital tissue image associated with an image of osteolysis of the underlying bone and taking the contract moderately. There was no endocranial extension. The preoperative workup was unremarkable apart from the positive serology of the HbS antigen. Total tumor resection was performed with a resection margin extended to 4 cm from the tumor boundary on 09/16/2010, followed by a second operation on 12/01/2010, which consisted of skin grafting thin sample taken from the inner side of the right thigh (Figure 6). The patient was seen at 1 year, 2 years and 7 years postoperatively and there was no sign of recurrence (Figure 7).

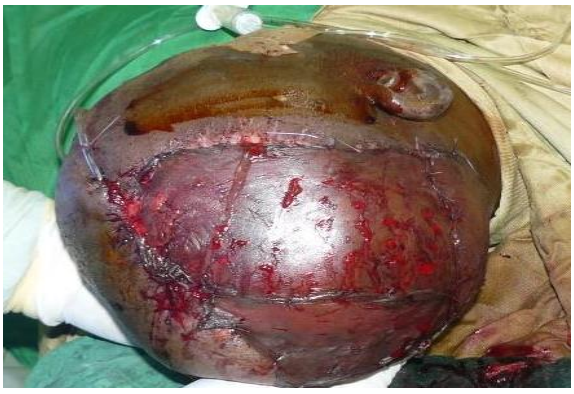

Fig-6: Appearance during the operation

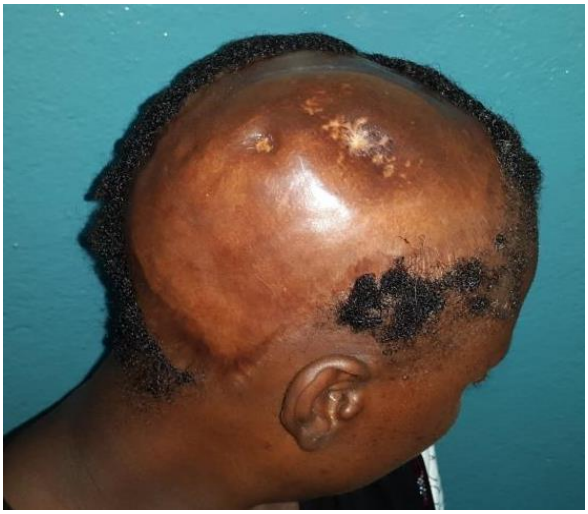

Fig-7: Postoperative aspect after 7 years

DISCUSSION

Darier and Ferrand's DFS or protuberant dermatofibrosarcoma is a rare mesenchymal tumor of intermediate malignancy. It represents less than 2% of all soft tissue sarcomas with an estimated incidence of around 4 cases per million [3-7]. The number of reported cases confirms this trend observed in the literature. In our series, all cases were female. On the other hand, some authors have reported a slight male predominance [8, 9]. It is generally found in adult subjects. The age of onset is between 20 and 50 years with averages ranging between 28 and 47 years according to the authors [5, 9]. This trend was also observed in this series. All breeds can be affected by DFS and there is no familial predisposition [10]. Some authors have observed the occurrence of DFS after local trauma in 10 to 20% of cases [11]. Other authors mention different exogenous factors in the onset of the condition such as scars from burns, vaccination, radiotherapy, traumatic node, syphilitic lesions, iatrogenic or occupational arsenical keratosis lesions. Pregnancy has also been suggested by some authors as a period of faster growth of DFS [8, 12]. In the cases reported, these notions have not been found.

Diagnosis is often delayed due to the torpid and long-tolerated development of the tumor. Tumors become very large in the absence of treatment. They push back the surrounding tissue and adhere to the deep plane or ulcerate to become painful and bleeding [8, 13]. This same observation was observed in our series. The size of the lesion can be extremely variable, usually depending on the time to consultation. It is on average 1 to 5 cm up to 40 cm [8, 9, 14]. In the reported cases, the size of the tumors varied from 15 cm to 20 cm. It was above the average observed in the literature. This could be explained by the longer consultation time leading to a diagnostic delay observed in our context. Usually, most lesions remain stable or progress gradually for a period of time before accelerated growth is noticed. The average time between the onset of the lesion and the first request for treatment is generally 7 to 9 years [8, 9, 12, 13]. This observation was made in our study. Confirmation of the diagnosis is given by pathological examination and possibly supplemented by immunohistochemistry. In the cases reported, the pathological examination confirmed the clinically suggested diagnosis. The immunohistochemistry with the search for reactivity to CD34 + was not carried out in our context because we do not have the appropriate technical platform. Standard x-rays, soft tissue ultrasound, CT and MRI provide information on the deep invasion of the tumor. CT gives a better analysis of the bone structures so the appearance of the tumor is that of a mass of soft tissue of tissue density without particular specificity [15]. Maxillofacial CT was performed in all patients. There was no secondary location after completion of the extension assessment.

Surgery with wide excision is the only therapeutic means effective in eradicating the tumor and preventing recurrence. Conventional surgery necessarily involves performing large and deep excisions in order to reduce the risk of recurrence, the margin of which has changed over the years [16]. The traditional surgical protocol provides for a 3 to 5 cm wide excision of the visible or palpable edges of the lesion and in depth of all the subcutaneous tissue up to the muscular plane included. Surgical excision can often be based on anatomical and functional territories and aesthetic units. First-line Mohs micrographic surgery is recommended because of the difficulties in this region [17]. This technique aims to reduce the surgical margins while controlling all the edges of the lesion.

Systematic dissection in the tumor drainage area is not justified because dissemination is more often carried out by the hematogenous route than by lymphatic extension. The loss of substance caused after excisional surgery is of variable size and depends on the location of the tumor and the adjacent tissue structures. Complications can occur especially in neglected forms that have evolved for several years: death by invasion of the brain in the DFS of the scalp, ulceration, bleeding spontaneously or caused by repeated rubbing or by trauma [18]. The progressive risk of recurrence of DFS is mainly linked to the quality of the first excision. The possibilities of recovery with well-conducted primary surgery are significantly greater than those with salvage surgery. DFS is not a lymphophilic tumor. Histologically proven lymphatic extensions are of the order of 1% of all distant tumor extensions. No metastatic localization was observed in the reported cases. DFS is a tumor with a good prognosis because, despite local recurrences, the vital prognosis is only exceptionally committed [2]. A retrospective multicenter American epidemiological study found a 5-year survival rate of 99.2% [19]. The deterioration in general condition was a factor for the poor prognosis in our case.

CONCLUSION

DFS is a rare malignant skin tumor with slow local progression. The progressive risk of recurrence is mainly linked to the quality of the first excision. This tumor can often be life-threatening in relation to its destructive character locally and the risk of recurrence. The possibilities of recovery in the event of well-conducted primary surgery are significantly greater than those of salvage surgery. Early diagnosis ensures rapid patient care and therefore prevents mutilating resections, the repair of which is sometimes complex.

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