Case Reprot

Open Access

Gigantic Dermatofibrosarcoma Protuberans of the Scalp with Fibrosarcomatous Transformation

Badr M I Abdulrauf^{1*} Jaudah Al-Maghrabi²

¹Section of Plastic and Reconstructive Surgery, Department of Surgery King Faisal Specialist Hospital and Research Center Jeddah, Saudi Arabia

²Department of Pathology and Laboratory Medicine King Faisal Specialist Hospital and Research Center Jeddah, Saudi Arabia.

ABSTRACT

Here we are reporting this rare and unfortunate presentation of Dermatofibrosarcoma protuberans (DFSP) of the scalp. The patient was a 30 years old male from a remote area, with history of 4 previous surgical interventions. He was transferred directly to our surgical ICU with septicemia; anuria; severe anemia; and respiratory failure. Patient had a massive fungating infected tumor on his scalp. CT scan shown a subcutaneous scalp tumor with maximum diameter of 45 cm and adherence to occipital bone but no intracranial invasion. The tumor kept bleeding whenever the blood pressure maintained to optimum level. Patient had to be taken to the O.R joined with Neurosurgeon in attempt of surgical control of bleeding by elevating the tumor from its bed. Unfortunately, patient collapsed intra-operatively and despite extensive attempts of resuscitation by Anesthesia team, patient could not be salvaged. The final pathology was DFSP with malignant sarcomata's transformation. To the best of our knowledge this is the largest reported DFSP case in the Head and Neck region, in English literature.

*Corresponding author

Dr Badr M I Abdulruf MD, FRCSC., Consultant Plastic Aesthetic and Reconstructive Surgery KFSH&RC. PO Box 40047 MBC J-40 Jeddah 21499 Saudi Arabia.

Received: April 23, 2020; **Accepted:** April 27, 2020; **Published:** April 30, 2020

Introduction

DFSP is a relatively rare and locally aggressive cutaneous Sarcoma with an incidence of 4.2 per million. DFSP is most frequently diagnosed between the age of 30 to 50 and is more common in Blacks [1]. DFSP typically has an insidious onset and a tendency to recur following surgical excision. Head and neck DFSP is an even less common site compared to the trunk.

The differential diagnosis of disproportionately large tumors on the head is broad, including skin appendage tumors, gliomas, neurofibromas, squamous cell carcinoma, malignant melanoma, desmoid tumors, sarcoma, and vascular malformations.

Histologically, conventional DFSP is characterized by the proliferation of dermal uniform spindle cells that extend into deep subcutaneous tissue. Classically, it is composed of monotonous cells with hyperchromatic and elongated nuclei that are arranged in a storiform pattern. Surgical resection with wide safety margins is the gold standard treatment for DFSP; other modalities include radiotherapy and imatinib for advanced disease [1].

Case presentation

This 30-year-old male patient from Yemen was initially admitted to a peripheral hospital for the last 40 days. Unfortunately, the patient was transferred to our hospital with septicemia secondary to an infected scalp tumor, intubated, mechanically ventilated,

and on vasopressors. As per history taken from relatives, he had this tumor for over 15 years and has had 4 surgical resections in the past.

Upon admission blood pressure was 70/30mm Hg, heart rate 26/min, temperature of 38 C° There was a large fungating tumor on the scalp. Purulent material was coming out associated with recurrent bleeding from its surface (Figure 1A & B).

Figure 1A: (A and B) Patient's head with a huge fungating tumor

Figure 1B

Patient was very pale, cachectic a 103 nd anuric. Computed tomography (CT) scan showed a tumor size with a maximum diameter of 45 cm and the involvement of occipital bone, but no intracranial extension (Figure 2). Hemoglobin was 6.8 gm/dl, white blood cell count of 18000/ mm²

Figure 2: CT cut showing a large subcutaneous mass measuring about 45 cm i006E maximum dimension

The patient was kept mechanically ventilated, resuscitated with IV fluids and blood products; empiric antibiotics as per infectious disease specialists; vasopressors; methylprednisolone in addition to local tumor care with dressings and bedside hemostasis. Patient basically kept bleeding from tumor surface at various sites while attempting to maintain the mean arterial pressure of 80mm Hg. Multiple attempts were made for bed side hemostasis with ligatures and fibrin sealing agents, however bleeding kept recurring. It was evident that hemostasis cannot be achieved unless the source is controlled in the tumor's bed.

A multidisciplinary meeting was conducted, and the patient's family was informed of the high risk of mortality in such conditions.

A day post admission a high-risk consent was obtained from the patient's next of kin and relatives in an attempt to surgical hemostasis with possible tumor debridement/resection. Neurosurgery were consulted and attended the surgery. The plan was that patient would be considered for a free microvascular flap such as latissimus dorsi muscle reconstruction of the defect at a later date.

Patient was shifted to OR while he was on significant hemodynamic support. Despite receiving several units of blood, his Hemoglobin was 5.1gm/dl. Soon after the tumor was elevated from its bed, unfortunately, the patient developed severe hypotension. Maximum resuscitation for approximately 45 minutes was attempted by the anesthesia team, but patient unfortunately could not be saved.

Gross pathological examination revealed a large multinodular tumor covered by skin and measuring 43 × 23 × 11 cm. Serial sections revealed a grey white to the yellowish cut surface with a firm consistency and associated with focal areas of myxoid and gelatinous appearance. Areas of necrosis were grossly identified. There was an area of necrosis covered by the tumor that showed a storiform pattern and revealed a low mitotic rate (2/10HPF). In focal areas, the tumor revealed sarcomatous transformation in the form of hypercellular areas with a Herringbone pattern and increased mitotic rate (5/10HPF) associated with necrosis. Immunohistochemistry assays revealed that the tumor cells are strongly positive for CD34 and vimentin. They were negative for smooth muscle actin (SMA), muscle-specific actin (MSA), S100, desmin, and myogenin. The areas with sarcomatous transformation show weak to absent CD34 staining. The final pathology diagnosis was a DFSP with fibrosarcomatous transformation (Figure 3).

Figure 3: (A) Hematoxylin and eosin-stained sections revealed a spindle cell tumor with a storiform pattern and involving dermis. (B) Immunohistochemical stain for CD34 shows strong positive staining in the classic DFSP area (upper left) and loss of staining in the fibrosarcomatous area (lower right)

Discussion

DSFP is well known to be only locally aggressive and rarely metastasizes, which explains the fact our patient had it for at least 15 years. Surgical resection with wide safety margins is the standard treatment; this patient has past history of four surgical resections, but the DSFP kept recurring. The recurrences tend to be higher in the head and neck for the obvious reason that operators tend to take suboptimal margins. Hence, Mohs micrographic surgery is commonly undertaken for DFSP in such areas. [1].

This patient spent less than 48 h in our hospital and was managed appropriately as a critically sick patient in the ICU. His condition was unstable; therefore, no further radiologic imaging of the tumor or staging was considered other than a chest x-ray, which was unremarkable. The decision to take him to the OR was made because, without intervention, he was expected to succumb to profuse hemorrhage directly from the tumor surface in the ICU. Interventional radiologic embolization was not an option due to patient's instability. Relatively large size tumors, including DFSP, have been previously reported; however, in terms of dimensions, we believe this is the largest reported DFSP case in the head and neck region. Sarcomatous transformation in DFSP is a rare phenomenon [2, 3]. It usually occurs in the form of fibrosarcoma and might appear as a well-demarcated area with uniform fascicular proliferation; Herringbone appearance, and

with an increased mitotic activity. However, in some cases, the sarcomatous transformation might resemble undifferentiated pleomorphic sarcoma/Malignant Fibrous Histiocytoma (MFH). Sarcomatous transformation in DFSP, either to Fibrosarcoma or MFH, is associated with an increased risk of recurrence and metastasis. Prognostic factors and survival in DFSP have been investigated [4]. Male patients and large tumor size (as in our case) at presentation are factors associated with poor prognosis and mortality.

References

1. Bogucki B, Neuhaus I, Hurst EA (2012) Dermatofibrosarcoma protuberans: a review of the literature. *Dermatol Surg* 38: 537-551.
2. Swaby MG, Evans HL, Fletcher CDM, Prieto VG, Patel KU, Lev DC, et al. (2011) Dermatofibrosarcoma protuberans with unusual sarcomatous transformation: a series of 4 cases with molecular confirmation. *Am J Dermatopath* 33: 354-360.
3. Szollosi Z, Nemes Z (2005) Transformed dermatofibrosarcoma protuberans: a clinicopathological study of eight cases. *J Clin Pathol* 58: 751-756.
4. Criscito MC, Martires KJ, Stein JA (2016) Prognostic factors, treatment, and survival in dermatofibrosarcoma protuberans. *JAMA Dermatol* 152: 1365-1371.

Copyright: ©2020 Badr Abdulrauf. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.