



Case Report

Trapezius Flap Reconstruction of Scalp Defect After Removal of Occipital Fibrosarcoma in Neurofibromatosis Type I Patient

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Abstract: Neurofibromatosis type 1 (NF1) is an autosomal dominant condition affecting approximately 1 in 3000 live births. The manifestations of this condition are extremely variable, even within families, and genetic counseling is consequently difficult with regard to prognosis. Individuals with NF1 are acknowledged to be at increased risk of malignancy. Several studies have previously attempted to quantify this risk, but have involved relatively small study populations. Soft tissue tumors represent a heterogeneous group of mesenchymal and neural lesions. We report a case of giant scalp Fibrosarcoma of the scalp in patient with neurofibromatosis type I without intracranial extension, in a 35 year old female which was excised completely along with the involved overlying skin, and reconstruction was done to cover the defect using trapezius flap and split thickness skin graft from the right thigh. She is doing well after treatment and is in regular follow up while awaiting further management by the oncologists.

Keywords: Neurofibromatosis Type I, Fibrosarcoma, Trapezius Flap, Scalp Defect

1. Introduction

Soft tissue sarcomas are a group of heterogeneous tumors that have their origin primarily in the embryonic mesoderm; more than 50 histological subtypes and diverse clinical behaviors have been identified [1]. Soft-tissue sarcomas are relatively rare in the head and neck. They account for 4–15% of all soft-tissue sarcomas and less than 10% of all neoplasms at this site. However, it is necessary to consider sarcoma in the differential diagnosis in order to avoid serious errors in

treatment. [2]. Sarcomas occur more commonly within several hereditary cancer syndromes, including retinoblastoma, Li-Fraumeni syndrome, neurofibromatosis type I, and familial adenomatous polyposis [3].

Fibrosarcoma is defined as a malignant spindle cell tumor that shows a herringbone or interlacing fascicular pattern without the expression of other connective tissue cell markers [Sapp JP et al, 2004]. Fibrosarcoma can arise in soft tissues or within bones.

The reconstructive options for a posterior scalp defect can usually be determined on the basis of the size of the wound.

Small defects (<2 cm²) can be closed primarily, medium defects (2–25 cm²) may be reconstructed using rotation-advancement flaps, and large defects (>25 cm²) can be reconstructed with larger rotation flaps [4, 5]

Pedicled or free tissue transfers may be required to provide coverage for large defects as well. Some of the notable options include a latissimus dorsi free flap, a pedicled trapezius myocutaneous flap, an anterolateral thigh flap, or an omental flap [6, 7].

2. Case Presentation

A 35 year old female presented with a large, gradually increasing swelling in occipital region since 18 years. The mass has been gradually growing in size then 3 months prior to our consultation the mass burst and started producing blood stained and smelling discharge. She had no family history of same conditions.

A complete physical examination was performed which revealed a fungating occipital mass, measuring 25x18x22cm in diameter, irregular in shape, brown in color, firm, fixed to the overlying skin, non-pulsatile; painless skin nodules over the trunk and extremities, suggestive of neurofibromas; hyperpigmented, irregularly shaped macules over the chest and back, suggestive of cafe-au-lait spots, twelve of which were larger than 15 mm; and both axillary and inguinal freckling were present. The mucous membranes were not affected. These findings were consistent with the diagnosis of neurofibromatosis type 1. His vital signs were within normal limits, The full blood count showed anemia with hemoglobin of 7.4 g/dl (14-18g/dl); Urea, creatinin and electrolytes were normal.

The lesion was sent for histology which showed a cellular lesion comprised of bundles of spindle cells with scanty eosinophilic cytoplasm, arranged in interlacing fascicles to produce a herringbone pattern. Focal necrosis was seen and mitoses averaged 4/10hpf. In keeping with a

Fibrosarcoma Grade II. The patient was taken to theatre and wide (2cm) margin excision was done, the mass was found to be very vascular. The patient had a cardiac arrest just after total excision of the mass and resuscitation was successful. Defect was left on the occiput with exposed bone. The patient was put on antibiotics and analgesia and was transfused then transferred to plastic surgeons for scalp defect reconstruction.

The tumor immunoreacted negatively for CD34, S100, SMA and Desmin. A month and half latter the patient was taken to theatre by plastic surgeons for trapezius flap to cover the defect, the flap was rotated to cover the exposed skull and the skin graft from the right thigh was used to cover the rest of the scalp defect with granulation tissue. One day after reconstruction power on the right upper limb was 2/5 and returned to normal on day 5 post operation but she could raise the limb above 90° with some difficulties and on day 15 after trapezius flap reconstruction the patient was discharged with a transfer letter to oncologist.

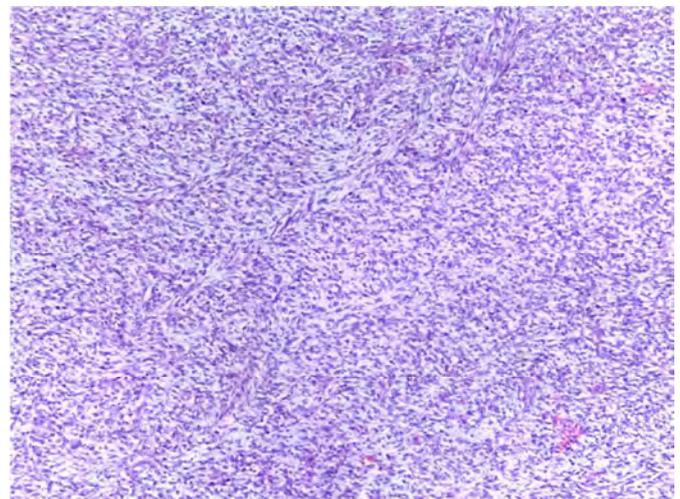


Figure 1. x20 magnification. A cellular lesion comprised of fascicles of spindle cells arranged in interlacing fascicles to produce a "herringbone" pattern. Vessels are seen between the fascicle.

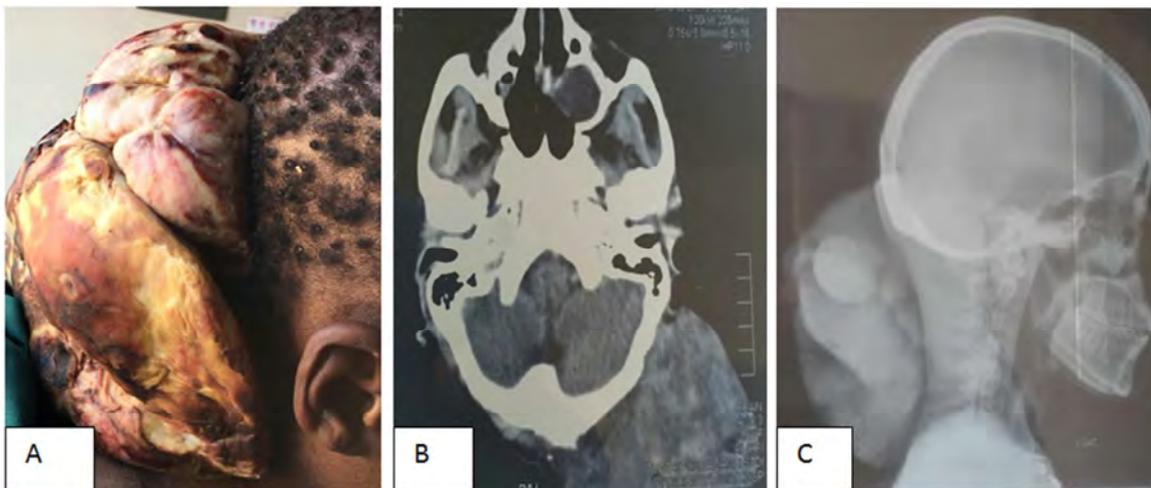




Figure 2. A. Picture of the Patient on Presentation Day. B. CT Scan Brain Showing a Giant Mixed Dense Soft Tissue Mass at Occiput Adhering to the Occipital Muscles with Multiple Free Gas Noted in it, no Erosion of Skull Bone, no Abnormality is Revealed in Intracranial Brain, There is Fluid Filling in Bilateral Maxillary Sinus. On Post Contrast CT Images the Mass Demonstrates Non Homogeneous Prominent Enhancement. C. X-ray of Head and Neck Lateral View. D. Preoperative View of the Defect. E. postoperative View Showing the Trapezius Flap and Partial-thickness Skin graft from the Thigh Applied to the Rest of the Scalp with Granulation Tissue. F. Appearance on Day 5 postop. G. Final Appearance on Day 15 Postop.

3. Discussion

Head/neck sarcomas are rare, accounting for about 1% of head/neck malignancies and 5% of sarcomas. Outcomes have historically been worse in this group, due to anatomic constraints leading to difficulty in completely excising tumors, with high rates of local recurrence [8, 9].

A prospective study of neurofibromatosis type 1 cancer incidence in the United Kingdom reported a preponderance of brain/CNS and connective tissue tumors, whereas although sarcomas were reported in 7% of individuals with NF1 [10]. Previous reports of malignancy rates in NF1 had varied in their assessment of the risks associated with this condition. In the longitudinal study by Sorensen et al (1986), 212 patients with NF1 were followed up over a period of 40 years, and 57 malignant tumors were found in this population [11].

Another longitudinal Scandinavian study [Zoller et al, 1997] estimated the rates of malignancy in 70 adult patients with NF1. Around 15% of patients in this cohort developed malignant tumours in a 12-year follow-up period [12]. Adult

fibrosarcoma is a malignant tumour, composed of fibroblasts with variable collagen production and, in classical cases, herringbone architecture. It is distinguished from infantile fibrosarcoma and from other specific types of fibroblastic sarcomas [13].

The head and neck sarcomas include a wide variety of histologic types of malignancy. Generous biopsy and accurate anatomic localization by CT are necessary prior to surgical intervention, radiotherapy and/or chemotherapy. The clinical behavior of fibrosarcoma is characterized by a high local recurrence rate and a low incidence of local regional lymph node and/or distant hematogenous metastases. However, hematogeneous metastases may involve the lungs, mediastinum, abdominal cavity and bone [14].

The histological appearance of fibrosarcoma does not allow a distinction between a primary tumor of bone from one arising in soft tissue [Chen Y et al, 2007]. Histologically, the degree of differentiation is variable, from a low grade one resembling fibroma to a highly anaplastic tumor, thus presenting a diagnostic dilemma to histopathologists. [15.]

Fibrosarcoma can be graded as either a low or high grade of

malignancy. Low-grade fibrosarcoma shows spindle cells arranged in fascicles with low to moderate cellularity and a herringbone appearance as shown in our case. This type of fibrosarcoma has a mild degree of nuclear pleomorphism and rare mitosis, with a collagenous stroma. High-grade lesions show intense nuclear pleomorphism, greater cellularity and atypical mitosis. The nuclei can be spindle shaped, oval or round. The histological appearance of high-grade fibrosarcoma may be similar to other tumors, such as malignant fibrous histiocytoma, or synovial sarcoma. The positive immunostaining for vimentin, together with negative staining for muscular immunomarkers, helps to confirm a diagnosis of Fibrosarcoma. [16]

Surgery remains the mainstay of treatment for head and neck sarcomas. Noteworthy exceptions to this principle include most rhabdomyosarcomas and Ewing's sarcomas. Radiation therapy is indicated after resection of all high grade sarcomas, large tumors, and when margins of resection are close or microscopically involved. Adjuvant treatment with both radiotherapy and chemotherapy are often recommended in selected patients in an attempt to achieve local control and prevent distant disease. Indications are microscopic or gross positive margins of resection and narrow margins with large high-grade tumors.

Treatment Options for Head and Neck Sarcomas: Cosmetic and functional results are important in head and neck sarcomas and microscopic disease can often be controlled with radiotherapy, avoiding a radical resection. The role of adjuvant radiation therapy is to sterilize microscopic extensions beyond the resection margins. Radiotherapy improves local control and survival over results achievable with surgery alone. Chemotherapy is not effective as a single-modality treatment but may help control micrometastatic disease in the lungs and augment the local control provided by radiation therapy. Unfortunately, even aggressive multimodality therapy may fail to control primary disease. Local control and distant metastasis remain a therapeutic challenge for improving survival in high-grade tumors. [17]

The trapezius muscle is superficial, flat, triangular, and approximately 34×18 cm in size; it covers the posterior cervical and interscapular thoracic areas. Along with the pectoralis and rotator cuff muscles, the trapezius suspends the pectoral girdle from its muscular origin located along the external occipital protuberance, the medial third of the superior nuchal line, the ligament nuchae, and the spinous processes of the seventh cervical and twelfth thoracic vertebrae [18]. According to the classification of Mathes and Nahai [19], the trapezius muscle has a type II vascular pattern with dominant and additional minor vascular pedicles. The transverse cervical artery with its branches is the major feeding artery. The additional minor vascular pedicles are the occipital artery and posterior intercostal artery. [20]

Despite this, standard cadaveric studies have shown great vascular variability where both the TCA and the dorsal scapular artery have been shown to be the dominant pedicle and may explain the flap's unpredictable results. [21, 22]

While several variations in flap design have been described in the literature, the major flap options include the superior, lateral, and lower trapezius myocutaneous flaps. [23]

The superior flap can be used for coverage of the posterolateral neck and is especially valuable after radical neck dissection and to cover irradiated wounds. The lateral flap is used for external defects of the lateral and anterior neck as well as mucosal defects of the pharynx and the oral cavity. The lower trapezius myocutaneous flap, as utilized in our case, has the most versatility in clinical application and has the most use for reconstruction of the occiput. [22].

4. Conclusion

Patients with head and neck sarcomas should undergo wide excision with emphasis on removal of gross disease and attaining clear surgical margins. In most patients, except those with small, low grade lesions, postoperative radiation therapy should be added to maximize local control. Head and neck sarcomas are rare tumors that can present management difficulties. These tumors are best managed in a multi-disciplinary setting.

Defects resulting from bulky tumor resection of the head and neck region represent a reconstructive challenge. The trapezius musculocutaneous paddle flap based on the transverse cervical vessels is a useful tool for the reconstructive surgeon.

The knowledge of the variance of its anatomical vasculature is essential in the successful use of the trapezius musculocutaneous flap

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