



FIBROSARCOMA SCALP IN A NIGERIAN LADY. CASE REPORT

Neurosurgery

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ABSTRACT

Fibrosarcoma is a malignant tumor of mesenchymal cell origin. It arises from pathologically transformed spindle shaped fibroblasts with high mitotic rate. As per WHO classification of soft tissue sarcomas, fibrosarcoma is defined as part of the fibroblastic/myofibroblastic sarcomas. There are two types of fibrosarcoma: the infantile/congenital-type, and the adult-type. WHO has defined infantile type as an intermediate malignant rarely metastasizing tumour while fibrosarcoma occurring in adults is classified as a highly malignant tumor. Fibrosarcoma scalp is a very rare occurrence^{2,7}.

KEYWORDS

Head, neck, sarcoma, scalp tumor

CASE REPORT

A 35 years Nigerian lady presented with ulcerative mass lesion of scalp and upper back of neck (Fig 1,2) associated with neck pain & headache for a few months. She was anaemic, had no lymphadenopathy, no lump in any other part of the body. She had undergone excision of scalp lesion twice in the past, in year 2014 & 2016 at Saudi Arabia. Previous CT/MRI Brain films were not available but her histopathology report was Spindle cell sarcoma and immunohistochemistry showed Vimentin & CD34 positive in tumor cells, SMA & Desmin, S-100 were negative. Ultrasound abdomen was normal. CT head done at our institute, showed irregular shape mass lesion in occipital, parietal and posterior subcutaneous tissue of neck at the level of C1 vertebra, with bony erosion of parietal bone and had no intracranial involvement of brain parenchyma (Fig 3, 4, 5). Her hemoglobin level was 6.2 gm%, so multiple blood transfusions given prior to surgery. On first stage of the operation, along with Plastic surgeon, entire tumor of the scalp was excised en-bloc with 2 cm healthy margins deep to the pericranium, midline parietal bone was found eroded and tumor was adherent to the dura (Fig. 6, 7, 8). Tumor was shaved off from the dura and Collagen sheet dressing applied. On second stage of operation, multiple burr holes made on the exposed cortical bone of skull, without breaching inner table (Fig. 9); so as have bone marrow granulation growth to facilitate skin grafting. Patient developed good bone marrow granulation over the cranium. On third stage of operation, our Plastic surgeon team, had harvested split skin graft from patient's thigh and applied it over the granulating cranium. There was good healthy take up of the graft (Fig. 10). Patient was discharged and followed up for 6 months, during that period there was no local recurrence.

HPE Report – Well defined cellular tumor involving subcutaneous fat composed of intersecting fascicles and bundles of spindle cells. Interspersed collagen bands present. There is minimal cytologic atypia. Mitosis is 3-4/10 HPF. Prominent staghorn vessels seen. Dense neutrophilic infiltrate with suppurative necrosis seen focally.

IHC panel: Vimentin, CD34, Bcl2 were positive, EMA, CK, SMA, Desmin, S100, CD99 & TLE1 were found negative.

Ki-67: 6-8%

Histomorphology and IHC findings favour solitary fibrous tumor.



Fig. 1

Fig. 2

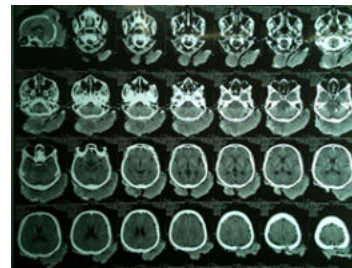


Fig. 3 (NCCT Head: Soft tissue tumor of scalp & neck)

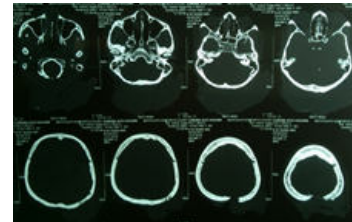


Fig. 4 (Bone window, revealing bony erosion)

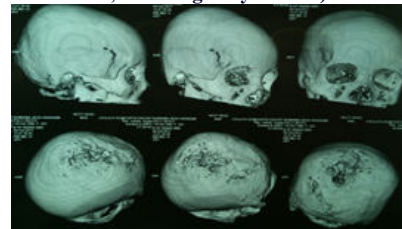


Fig. 5 (Erosion of parietal bone)



Fig. 6 (Intraoperative view)

Fig. 7

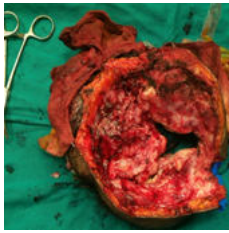


Fig.8(Excised specimen)

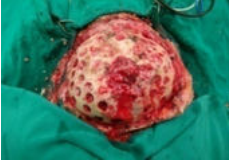


Fig.9 (Multiple burr holes on cranium)



Fig. 10 (Split skin graft applied over granulating cranium)

DISCUSSION

Fibrosarcoma arises in regions consisting of collagen-rich connective tissue. Adult-type fibrosarcoma of head or neck region is an exceptionally rare clinical entity^{2,7,8,9}. Management of fibrosarcoma scalp, requires a multidisciplinary approach (Neurosurgeon, Head & neck surgeon, Plastic surgeon, Radiation-oncologist). Surgical en-bloc resection is the standard modality of treatment for the localized soft tissue sarcomas. A 2 cm tumor free margin of healthy tissue resection (R0) is recommended, so as to minimize the risk of local recurrences⁸. This means that not only the tumour tissue itself, but also part of the adjacent healthy tissue has to be removed due to the infiltrative growth of fibrosarcoma, in such cases post-operative radiation therapy is not recommended^{10,11}. Lesions which are deep seated, high-grade, > 5 cm size, radiation therapy after an R0 resection is highly recommended^{1,12,13}. Reoperation must be considered in the case of R1 resections (microscopic tumor at the margin) and also in the case of R2 surgery (macroscopic tumor at the margin), reoperation is mandatory^{1,12}. Radiotherapy will follow R1, R2 excisions, if these cannot be rescued through re-excision¹. Adjuvant chemotherapy in soft tissue sarcomas is very controversial¹².

This case was managed with multidisciplinary team (Neurosurgeon, Plastic Surgeon) and achieved successful multiple staged operations, with no recurrence in 6 months follow up.

CONCLUSION

The best therapy for fibrosarcoma is en-bloc surgical removal and requires multidisciplinary team engagement, for its better management. Even though the response rate of fibrosarcoma towards radiotherapy and chemotherapy is very low, they are broadly used as a neoadjuvant and/or adjuvant tumor treatment^{1,7,12}.

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