# Infantile fibrosarcoma of the parotid gland in a 6-year old female: case report and management challenges

Case Report
**Evaristus E AFIADIGWE**[1]
**Basil C EZEANOLU**E[1]
**Cornelius C UKAH**[2]
**Victor I ONYIAORAH**[2]
**Titus O CHUKWUANUKWU**[3]
**Thomas O ULASI**[4]

[1] *ENT Unit*
[2] *Histopathology Department*
[3] *Plastic Surgery Unit*
[4] *Paediatric Oncology Unit*
*Nnamdi Azikiwe University*
*Teaching Hospital*
*Nnewi*
*Anambra State, Nigeria*

***Author for Correspondence***
**Dr Evaristus AE Afiadigwe**
ENT Unit
Department of Surgery
Faculty of Medicine
College of Health Sciences
Nnamdi Azikiwe University
Nnewi Campus
Anambra State, Nigeria

**Email:**
drevafiadigwe@ymail.com
Phone: 08034059292

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Infantile fibrosarcoma is a rare soft tissue sarcoma that may present at any time from birth to early childhood. The parotid gland is not a common site for malignancies especially in children, and there has been no report of this disease in our environment.

The aim of this publication is to report a case of this rare tumour, managed recently in our Centre, and to highlight some of the attendant challenges during the management.

We present the case of a 6-year old female child with infantile fibrosarcoma of the left parotid gland and subtle left facial palsy – probably the first in Nigeria; emphasizing the rarity of this tumour, and the need for a high index of suspicion in making the diagnosis.

***Keywords:*** *Chemotherapy, facial palsy, infantile fibrosarcoma, parotidectomy*

ABSTRACT

## INTRODUCTION

Infantile fibrosarcoma (IF) is a rare type of soft tissue sarcoma which could be seen at birth or in early childhood. It comprises less than 1% of all childhood tumours and approximately 10% of all sarcomas.[1] Tumours most commonly present at the extremities (61%), trunk (19%), and head / neck (16%).[2]

In the general population, majority of parotid tumours are benign, and so, malignant tumours in paediatric age group is most uncommon. Although histologically similar to the adult fibrosarcoma, infantile fibrosarcoma clinically differs in local recurrence, with rare distant metastasis and generally, better prognosis.[3,4] The authors are not aware of any report of infantile fibrosarcoma of the parotid gland which has been published from this country.

Wide surgical excision is the mainstay of treatment, although it is also known to be a chemo-sensitive tumour and as such,

adjuvant or neo-adjuvant chemotherapy is also employed in its management, especially where a wide excision is difficult to achieve.[4,5]

CASE REPORT

A 6-year old female child presented to our Ear, Nose and Throat (ENT) Clinic with a lump below the left ear of 3months' duration. The mass progressively increased in size after an incisional biopsy prior to the referral. There were no complaints of pain, facial weakness, throat ulceration, dysphagia, odynophagia, otalgia or change in appetite, and there were no other lumps in the head and neck region.

Physical examination revealed a left parotid mass with irregular surface, 6cm x 8cm in diameter, non-tender, firm to hard in consistency and immobile, but not attached to the overlying skin. An incisional scar and mild left ipsilateral facial palsy were noted but there was no cervical lymphadenopathy. Left tonsillomegaly was also noted but no other features of malignancy.

**Figure 1:** Left Parotid tumour lobulated

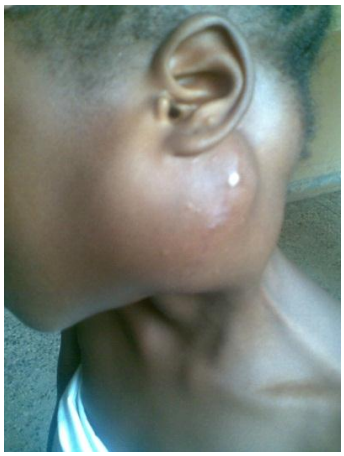
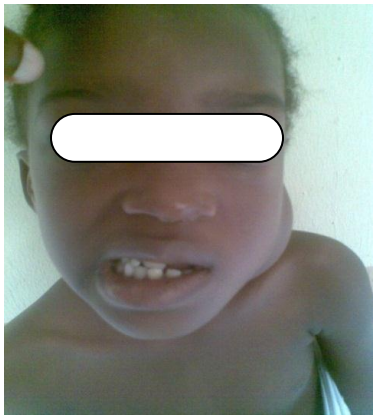


**Figure 2**: Front view showing left facial palsy



Laboratory investigations including blood count, urinalysis and human immune deficiency virus screening, were all normal. Fine Needle Aspiration Cytology (FNAC) was reported as 'Suspicious, probably Benign'. Subsequently, Examination Under Anaesthesia (EUA), biopsy tonsillectomy and parapharyngeal space incision biopsy were performed, and the histology report read:

> *Tonsils – reactive nodal hyperplasia; left parapharyngeal specimen - well circumscribed benign neoplastic mass consisting of proliferating fibroblasts - fibroma.*

On the basis of the above findings, a provisional diagnosis of a benign left parotid tumour ?malignant, was made and so, the patient was worked up for parotidectomy.

Total parotidectomy was done with in-continuity dissection of the occipital node extension. Outcome was good with residual facial palsy. The parotid specimen was subjected to histology and was then reported as infantile fibrosarcoma.

**Figure 3:** Lateral view 5th day post-op

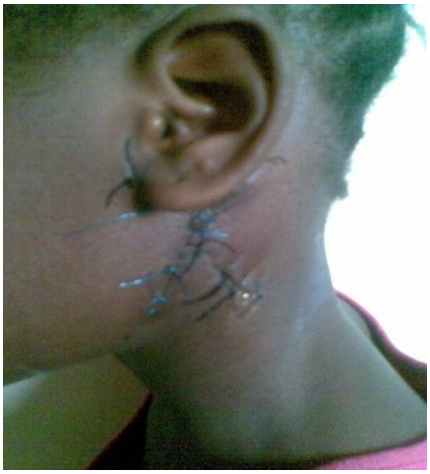
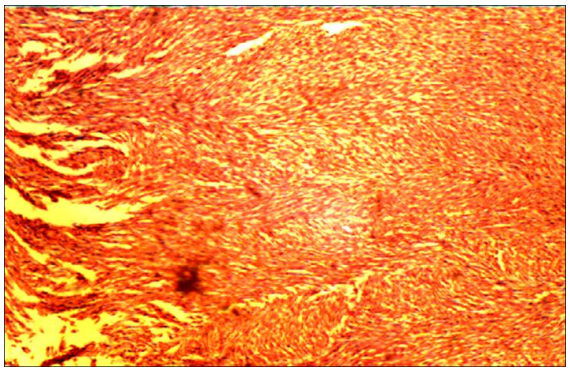


**Figure 4:** Photomicrograph X100 magnification, showing proliferating spindle-shaped cells with no native parotid gland seen due to a complete effacement by these neoplastic cells



On account of the high recurrence rate associated with infantile fibrosarcoma (IF), the patient was commenced on chemotherapy using the most common regimen consisting of vincristine, actinomycin-D and cyclophosphamide (VAC).[3]

## DISCUSSION

Generally, malignancies are uncommon in the parotid gland especially in childhood as most parotid tumours are benign. It is, therefore, understandable that we thought we were dealing with a benign lesion or that the primary lesion was in the left tonsil. But, we were wrong! The apparent left tonsillar enlargement was most likely due to its displacement by the deep parotid lobe.

Infantile fibrosarcoma is histologically similar to the adult fibrosarcoma but its clinical manifestations are quite different. Unlike the adult variety, the congenital or infantile fibrosarcoma is associated with local recurrence but metastasis is uncommon giving a generally better prognosis.[4]

The treatment of choice is wide resection around the tumour, and in the extremities this may extend to the outright amputation of a limb. Where possible, total excision is the gold standard and can even be curative.[5] Considering the peculiar anatomy of the head and neck, wide excision is difficult and sometimes inconceivable, and so, conservative surgery and chemo/radiotherapy are recommended.

The role of chemotherapy either alone or in combination with surgical resection has been well documented[3-10]. Neo-adjuvant chemotherapy has been shown to shrink the tumour considerably, and by so doing, converts an inoperable lesion to an operable one. Drugs effective in this include ifosfamide, doxorubicin, etoposide and adriamycin. The most commonly used combination is vincristine, actinomycin-D and cyclophosphamide (VAC), which was the regimen used for the index patient.[9-12] This regimen has been found effective in several studies even as the only successful treatment regimen.[10]

The cost and/or unavailability of chemotherapy and radiotherapy was/were the major hindrance in the care of this patient in our resource-constrained country. She would require a long term follow up in the expectation that she will comply. Another challenge envisaged in the management of this patient is, follow up. In our society long

term follow up remains an issue no thanks to ignorance and poverty[13].

CONCLUSION

Infantile fibrosarcoma is a rare disease, and so, a high index of suspicion is required for early diagnosis. There is need to equip our centres with relevant facilities if the fight against cancer is to be won, since cancer management requires a multi-disciplinary approach.

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