**CASE REPORT**

**Congenital infantile fibrosarcoma mimicking sacrococcygeal teratoma in a Ghanaian infant: A case report and review of the literature**

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Congenital infantile fibrosarcoma (CIFS) is a rare tumour of childhood accounting for less than 1% of malignant tumours in children. It accounts for <1% of malignant tumours in children.1,2 The varied pattern of presentation of this cancer can be deduced from the occasional case reports that have emanated from almost every continent.2-5 Reports from sub-Saharan Africa are particularly rare and the occurrence in the sacral region mimicking a sacrococcygeal teratoma has been reported,2-5 and such presentation is generally uncommon. The poor predilection of the tumour for distant metastases may contribute to its above average prognosis, but the rarity of systemic antenatal detection of congenital conditions in most parts of Africa may mitigate this good fortune. We report the very rare case of a 9-month-old female infant who presented with a progressively increasing painless sacral mass from birth which was thought to be a sacrococcygeal teratoma clinically but histopathological assessment revealed a CIFS.

**Case report**

A 9-month-old female infant was brought to our outpatient department by a distraught mother. The infant was born with a painless sacral mass, which had been progressively increasing in size. Her mother had sought help from faith healers and traditionalists, which accounted for the late presentation. There were no associated local or systemic symptoms. There was no history of maternal illness during pregnancy nor was there use of unprescribed medications. Physical examination revealed a healthy-looking female infant who was not pale, no pedal oedema or significant lymphadenopathy. There was a 25 × 20 cm mass of mixed consistency overlying the sacral region, extending to both gluteal regions and partially surrounding the anus (Fig. 1). Anal sphincteric tone was found to be slightly reduced on digital rectal examination. A tentative diagnosis of sacrococcygeal teratoma. Pathological examination of the surgical specimen revealed a CIFS.

We report the case of a 9-month-old female infant thought to have a sacrococcygeal teratoma. Pathological examination of the surgical specimen revealed a CIFS.

Congenital infantile fibrosarcoma is a rare tumour of childhood. It occurs in any part of the body and has been found in unusual parts of the body,16 however, the extremities, namely the head, neck and trunk, in that order, are the most frequently affected locations.16 The lesions in the trunk are usually more aggressive and recurrence rates are high, especially when the excision is incomplete. Presentation in the sacral region has been reported by Al-Salem;3 this is the only reported occurrence in this location. Our patient presented in a similar fashion and we understandably made a misdiagnosis of sacrococcygeal teratoma. Sacrococcygeal teratomas present at birth as sacrococcygeal masses.10 Our curiosity was, however, aroused by normal serum alpha-fetoprotein levels. This tumour marker, although not diagnostic of sacrococcygeal teratomas, is elevated in many cases.12 The margins of resection were involved, which would have been avoided if a biopsy had been entertained following ultrasonography of the sacral mass.13 Infiltration of surrounding structures by the tumour is well documented. This open access article is distributed under Creative Commons licence CC-BY-NC 4.0. This open access article is distributed under Creative Commons licence CC-BY-NC 4.0.
was in keeping with the general principle of reducing the risk of recurrence since we had assumed it was a sacrococcygeal teratoma.\[13\]

Although cytogenetic analysis of CIFS has been shown to be useful in confirming diagnosis, revealing the fusion transcript ETV6-NTRK3, such studies are not routine in our centre. The biological behaviour of the tumour and its chemosensitivity have been attributed to this defect, which itself is the result of a recurrent chromosomal translocation t (12;15) (p13; q25).\[1,2,5\,7\] This is a translocation between the ETS-related transcription regulator chromosomal translocation t (12;15) (p13; q25).\[1,2,5,7\] This is a defect, which itself is the result of a recurrent chromosomal translocation t (12;15) (p13; q25).

Neo-adjuvant chemotherapy is the current treatment approach, which makes surgery less mutilating. Vincristine and actinomycin D are the most frequently employed agents.\[1,2,4\] CIFS has been successfully treated with chemotherapy alone.\[4,5\] Two percent of patients in the series reported by Orbach et al.\[4\] and 2 of the 11 in that by Lohl et al.\[2\] received radiotherapy in addition to surgery and chemotherapy. Our patient received vincristine and actinomycin D chemotherapy. The risk of recurrence is generally high, especially in fibrosarcoma of the trunk, and therefore our patient is periodically followed up after chemotherapy. Differential diagnoses to be entertained in masses located in this abnormal area in our patient should include, but not be limited to, embryonal rhabdomyosarcoma, neurofibroma and schwannoma.

**Conclusion**

CIFS can mimic a sacrococcygeal teratoma. The possibility of this diagnosis should be entertained especially if the serum alphafetoprotein levels are not elevated. Radiological imaging may not help with differentiation. In our opinion, a core biopsy in such instances may help determine the appropriate therapeutic approach, as CIFS is chemosensitive. Neo-adjuvant chemotherapy would allow for complete resection without sacrificing vital structures. With the reported excellent survival rates of >80%, the outlook for our patient is good.\[4,5\]

**References**


Fig. 1. A soft non-tender tissue mass on the sacrococcygeal area of the infant.

Fig. 2. Microscopy of the sacrococcygeal mass showing hypercellular tumour composed of plump spindle cells with elongated nuclei and abundant eosinophilic cytoplasm arranged in storiform and herring-bone pattern in areas. There is rare abnominal mitosis. (A, B, C and D are H&E stains at × 40, 100, 200 and 400, respectively.)