East African Medical Journal Vol. 79 No. 2 February 2002
FRONTO-ETHMOIDAL TERATOMA: CASE REPORT
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FRONTO-ETHMOIDAL TERATOMA: CASE REPORT

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SUMMARY

A one-month old baby was referred to the neurosurgical unit, Kenyatta National Hospital, Nairobi, Kenya, with history of being born with a bony outgrowth from the right side of her face which resembled an undeveloped twin. A computerised tomography scan of the brain and the extracranial mass confirmed a defect in the anterior cranial fossa and extension of the mass intracranially. Total surgical excision of the mass was done and facial reconstruction achieved. Histology of the excised mass confirmed a mature (benign) teratoma. Except for location, fronto-ethmoidal teratoma resembles its counterpart in the sacro-coccygeal area. It may arise over the nasion, or within the nose, orbit, or mouth and frequently extends intracranially. Like other teratomatous tumours, malignant changes tend to occur with increasing age. Calcification within the mass is often evident on plain skull x-rays. Treatment consists of early total excision.

INTRODUCTION

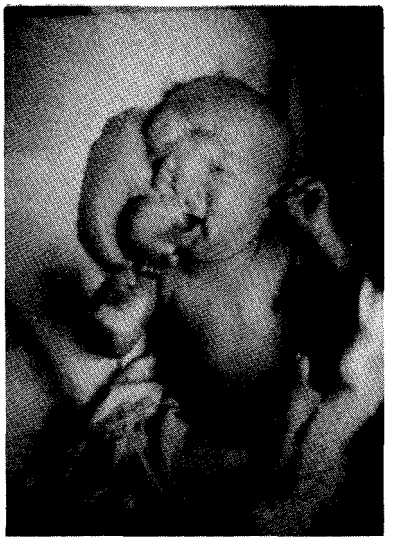
Cranial-facial teratomas are rare. They are classified as germ cell tumours. Germ cell tumours include germinoma, mature and immature teratoma, malignant teratoma, embryomal carcinoma, yolk sac tumour, choriocarcinoma, and mixed GCT'S. All these tumours originate from the primordial germ cell in the embryo(1). These cells arise from the embryonic yolk sac endoderm and migrate toward the gonadal folds. Germ cells that are topographically misplaced during migration do not survive and are eliminated by an immune mechanism(2). If they survive they may acquire neoplastic properties due to unknown oncogenic factors or mutant genes(3). Totipotent neoplastic germ cell may mature along embryonal or extra embryonal lines. Embryonal differentiation results in teratomatous elements. Extra embryonal differentiation results in formation of choriocarcinoma, yolk sac tumour and embryonal carcinoma(2).

CASE REPORT

A one month old female was referred to the Neurosurgical unit, Kenyatta National Hospital, from a peripheral District Hospital because of a cranio-facial malformation observed at birth. This was in the form of a bony outgrowth from the face with features resembling a mouth, eyes, nose and limbs, creating an impression of an undeveloped twin (Figure 1). She was a third born in a family of three from a single mother. The pregnancy was uneventful and it was a normal term delivery. A plain x-ray of the head showed features of long bone formation in one of the appendages. Computerised tomography (CT) scan of the brain showed a defect in the anterior cranial fossa with some intracranial extension of the mass. Neurological examination was normal. Systemic examination was normal. Routine investigations were normal. The child was prepared for surgery.

Figure 1

Fronto-ethmoidal teratoma: histology confirmed a benign teratoma



Intervention: The patient was given a general anaesthetic, with endotracheal intubation. Detailed examination of the malformation revealed a medial swelling with skeletal component and well formed soft tissues and skin. There was an inferior swelling arising from the nasal septum and nasal bridge, spherical and multiloculated. There were structures resembling mouth, nose and fingers on the superior aspect of the mass. There were some tufts of hair in some areas. There was an opening (sinus) on the right side of the nose communicating with the maxillary antrum.

A skin flap was raised from the right side and the multiloculated mass dissected out. The mass was cystic and covered by dura mater. The limb like appendage was dissected up to the base and resected. The dura was opened and the multiloculated cystic mass dissected free up to its base.

Haemostasis was achieved and the defect repaired with tight dural closure. Reconstruction of the face was done using the skin from the appendage. The patient tolerated the procedure well. Post-operative course was uneventful and she went home on the tenth post-operative day.

Histopathology report: Gross: Bulky fragments, some covered by skin ellipse (measuring 13mm in diameter). One fragment had a long bony finger-like structure covered by skin, another had a nipple-like structure. One fragment had a cyst containing sebaceous material on section.

Microscopy: Sections showed a lesion with cystic spaces, some lined by skin, ciliated columnar epithelium and colonic type mucosa. The features were in keeping with a mature (benign) teratoma.

DISCUSSION

Massive congenital intracranial teratomas, with extracranial extension are rare. The prognosis is usually poor with still birth or immediate post-partum death as the usual outcome. Craniofacial reconstruction is usually necessary after radical surgical resection in those patients who survive. A similar case to the one presented in this paper has been reported by Lanzino *et al*(4) with encouraging four year follow-up results. The histological similarity between seminomas and some pineal tumours (pinealomas) was first reported by Russell in 1944(5). She suggested that these tumours were actually teratoid tumours. Later, in 1946, Friedman and Moore(6) proposed that seminomas arose from primordial germ cells which they named germinomas and put forward the germ cell theory to explain the origin of these tumours. The germ cell theory proposed that germ cells (totipotent cell) gave rise to germinoma, embryonal carcinoma, choriocarcinoma and teratoma. This theory has received the WHO brain tumour classification committee approval(1). Germ cell tumours comprise germinomas (seminoma and dysgerminoma), mature and immature teratoma, malignant teratoma, embryonal carcinoma, yolk sac tumour, choriocarcimoma and mixed GCT'S(1).

Germ cell tumours (except for germinomas) arise by parthenogenetic fertilisation of the germ cell(7). The totipotent neoplastic germ cell may mature along either embryonal or extraembryonal lines. Embryonal differentiation results in the formation of teratomas, extraembryonal differentiation results in formation of choriocarcinoma, yolk sac tumour and embryonal carcinoma. There are two groups of teratomas(8). The first type seen in the gonads and posterior abdominal wall is derived from germ cells by a process of parthenogenesis. The second type seen in the sacro-coccygeal region, the head and the chest is related to sequestration of cells of the blastula before differential blocking of the genome has occurred and may be regarded as a derivative of an incomplete conjoined twin. An intracranial teratoma has two peaks in its age distribution. The first peak is in the neonatal/infancy period (10%) and the other peak is in the childhood period between 5-14 years. Teratoma in the first peak may be regarded as foetus-in-fetu and teratoma in the second peak derives from the germ cells.

No mechanism has been found to explain how the primordial germ cells are incorporated into the cranial cavity considering the fact that the route of their physiological migration is far from the cranial cavity. In this paper, a case of fronto-ethmoidal teratoma treated surgically with good results has been presented and discussed. Long term follow up is currently in process.

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