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Case Report

Congenital Infantile Fibrosarcoma of the Tongue: Case Report and Review of the Literature

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Abstract

Introduction: Congenital infantile fibrosarcoma is often misdiagnosed. It may be more common than noted in the literature. We present an unusual case of paediatric head and neck fibrosarcoma.

Methods: Restrospective case report and literature review.

Results: We report the surgical management of a fibrosarcoma of the tongue in an 8-month-old child where neo-adjuvant chemotherapy was unsuccessful.

Conclusion: Surgical resection is the mainstay of treatment, in the head and neck region where critical structures are close to the tumour, complete resection with wide margins can be difficult. The role of chemotherapy is yet to be defined.

Keywords: Fibrosarcoma, infantile, paediatrics, tongue, congenital, management

Introduction

Congenital infantile fibrosarcoma is an uncommon tumour that usually occurs in the first four years of life, especially in the first three months, in the deep soft tissue of the distal extremities with 29% presenting in axial locations [1,2]. Despite its' rarity, it is the most common soft tissue sarcoma in children below one year old and represents between 20% to 50% of the malignant soft tissue tumours in neonates [3]. As congenital infantile fibrosarcoma is a hypervascular

tumour, it is often misdiagnosed as the more common congenital haemangioma [4,5].

Surgical excision has remained the definitive management [1,6-9]. We report a rare case of fibrosarcoma involving only the tongue of an 8 month old child where surgical resection was performed to ensure clear margins and to preserve as much of the patient's tongue and surrounding structures as possible. Neo-adjuvant chemotherapy was attempted to optimise clinical outcomes but was unsuccessful.

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Case Report

A term male was born after an uncomplicated pregnancy. At birth, the infant was noted to have a left-sided lingual vascular lesion which was clinically and radiologically diagnosed to be a haemangioma. At first there were minimal symptoms of feeding difficulties and airway obstruction and the lesion was treated with oral and intra-lesional steroids. However, the lesion grew and caused intermittent airway obstruction. He was referred to our institution at two weeks of age and underwent a partial wedge resection of the anterolateral tongue lesion at two months of age after failure of conservative treatment.

The differential diagnosis of the frozen sections demonstrated spindle cell neoplasm with multiple mitoses, which was thought to be either infantile myofibromatosis or infantile fibrosarcoma. However, there were features favouring infiltrative myofibromatosis and also focal appearances of infantile fibrosarcoma, so a definitive diagnosis could not be made. Neo-adjuvant chemotherapy was offered to de-bulk the tumour prior to definitive surgical excision.

Four courses of the VAC (Vincristine, Actinomycin, Cyclophosphamide) chemotherapy were planned over a period of twelve weeks. The tumour size significantly reduced after the first course. However, as there was no further radiological improvement despite completing the four courses, the family agreed to proceed with surgical resection of the tumour.

At 8 months of age, the patient underwent subtotal glossectomy, floor of mouth resection, bilateral neck dissection and reconstruction with a full thickness skin graft. A tracheostomy tube was placed to assist with ventilation while a nasogastric tube was inserted for nutritional support (Figures 1-3).

With only just under a third of his tongue left, the patient underwent intensive speech therapy. At his three years follow up, he has a well articulated speech pattern and good oral function. At his four years follow up, there was no evidence of recurrence.

Discussion

Congenital infantile fibrosarcoma is an indolent, slow-growing but malignant spindle cell tumour [10]. Histologically, infantile fibrosarcoma exhibits spindle-shaped cells with the associated fibres arranged uniformly in interlacing bands or fascicles in a herring-bone pattern [11,12]. There are several different tumour gradings, with Smith et al. using the system devised for non-rhabdomyosarcomatous soft tissue sarcomas (NRSTS) by the Paediatric Oncology Group, where low-grade fibrosarcoma was defined as uniform spindle-shaped cells with few mitoses while high-grade fibrosarcoma was defined as spindle-shaped cells with pleomorphic features and numerous mitoses [13,14]. Mark et al. [15] followed the system of high or low grade fibrosarcoma according to the UCLA pathologists and Conley et al. [16] where low-grade fibrosarcoma is pathologically marked by sheets of uniform spindle cells with low to moderate cellularity arranged classically in a herring-bone pattern [17]. High grade lesions show a greater degree of nuclear pleomorphism, greater cellularity and more frequent mitotic activity with non-uniform spindle-shaped nuclei and presence of tumour necrosis [17,18]. In our case, the tumour was defined as a low-grade fibrosarcoma as it displayed uniform spindle-shaped cells with low cellularity and occasional pleomorphic nuclei.

Benign differential diagnoses of infantile fibrosarcoma include infantile haemangioma, juvenile xanthogranuloma, haemangiopericytoma, lipoblastoma, lipomas, encephaloceles, glioma, dermoid cysts, plexiform neurofibroma, Spitz naevus infantile myofibromatosis and orf infection. Malignant differentials to consider include dermatofibrosarcoma, giant cell fibroblastoma, rhabdomyosarcoma, lymphoblastic lymphoma, neuroblastoma, Langerhans cell histiocytosis, congenital leukaemia cutis, peripheral primitive neuroectodermal tumour and adrenocortical carcinoma [19].

The primary treatment of infantile fibrosarcoma is wide local excision and total resection to prevent any locoregional and distant recurrence. However, due to the propensity for extensive local invasion into surrounding tissue combined with the complexity of the

Figure 1: The left tongue following excision.

Figure 2: The post-operative appearance of the completed excision of left tongue fibrosarcoma.

Figure 3: The excised specimen.

anatomy of the head and neck region with associated critical structures, complete surgical resection is often difficult.

The role of chemotherapy pre-operatively or post-operatively in the management of fibrosarcoma in the head and neck region is controversial. Smith et al. [12] reported that despite receiving adjuvant chemotherapy, 4 of the 6 reported cases died of extensive local disease 8-9 months after the initial diagnosis. Mark et al. [15] reported a single case of adjuvant radiotherapy and chemotherapy with evidence of local disease despite treatment, while 3 of the other 8 patients required further surgical resection with mixed outcomes.

Despite a lack of consensus on the most appropriate post-operative therapy, the prognosis for patients with infantile fibrosarcoma is generally good. The overall 10 year survival for those presenting at less than two years of age is 80% compared with 60% for those with later onset presentations. Those with involvement of acral sites have a lower risk of metastasis (8%) and mortality (5%) than those with tumours involving axial sites (metastasis 26%; mortality 26%) [20]. Younger patients with acral lesions and those without evidence

of metastatic disease have the best prognosis [10].

A review of the literature since 1989 shows 22 children who were diagnosed with fibrosarcoma of the head and neck at various ages (Table 1). 72% (n=16) of the children were alive at the time of their respective publications, of which 81.25% (n=13) had no evidence of local disease, 12.5% (n=2) had local disease while 6.25% (n=1) had local disease with distant metastases.

The infantile fibrosarcomas were located in various locations in the head and neck region. 27.2% (n=6) involved only the mandible, 9.1% (n=2) involved only the maxilla, the posterior neck, skull base and the orbit. One case (4.5%) involved the skull base, cheek, scalp, tonsils, the posterior head, nasal cavity, hard palate, mandible, mastoid, tongue, hard palate and mandible. Two children had the tumour in multiple sites (Table 1).

The management was mainly wide excision or partial/subtotal/total resection of the tumour regardless of the site. Different degrees of resection were required due to the presence of surrounding vital structures of the head and neck. Six had wide excision, 9.1% (n=2) had partial excision, 40.9% (n=9) had subtotal resection

Table 1: Clinical features of reported cases of fibrosarcoma in the oral and maxillofacial region in children below 18 years old.

Year	Author	Age (years)	Sex	Site	Management	Status
1989	Bang et al. [21]	2.5	M	Mandible	Wide excision	ALD
1991	Mark et al. [15]	10	F	Skull base	Subtotal resection	ALD
		15mo	F	Nasal cavity, maxilla, orbit	Subtotal resection	ANED
		10	F	Scalp	Subtotal resection	ANED
		3	M	Mastoid, tongue, hard palate, mandible	Subtotal resection	DLD
		3	F	Orbit	Subtotal resection	ANED
		2	F	Cheek	Total resection	ANED
		10	F	Mastoid, brain	Subtotal resection	ALD
		16	M	Ear, cheek	Subtotal resection	DLD
		2	F	Orbit	Total resection	ANED
1995	Smith et al. [12]	12.5	F	Maxilla	Partial resection	DLD
		18	M	Maxilla	Total resection	DLD
		9	F	Mandible	Partial resection	DLDDM
		13	F	Tonsil	Wide excision	ANED
		9	F	Mandible	Wide excision	ANED
		7.5	M	Posterior neck	Subtotal resection	DLD
1998	Muzio et al. [22]	4	M	Mandible	Wide excision	ANED
2002	Loh et al. [23]	Birth	F	Posterior head	Subtotal resection	ANED
2004	Keller et al. [24]	8	F	Mandible	Wide excision	ANED
		2	F	Skull base	Total resection	ANED
2008	Gosau et al. [11]	9	F	Mandible	Wide excision	ANED
2008	Costa et al. [25]	3wks	M	Posterior neck	Total resection	ANED

A: alive, D: dead, NED: no evidence of disease, LD: local disease, DM: distant metastases

and 22.7% (n=5) had total resection.

Conclusion

In conclusion, congenital infantile fibrosarcoma may be more common than noted in the literature but is usually misdiagnosed. Tumour grade, size and adequacy of surgical margins are important prognostic factors in the management of infantile fibrosarcoma. While surgical resection is the mainstay of treatment, in circumstances where critical structures are close to the tumour, complete resection can be difficult. The role of chemotherapy is yet to be defined.

Conflict of Interests

None to declare.

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