# AN UNUSUALLY LARGE SPINDLE CELL SARCOMA OF THE TONGUE IN A MALE CHILD

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## **SUMMARY**

Reports of spindle cell sarcoma in the tongue are very rare, and seldom in the child. We report a five-year-old male patient who presented with a massive lesion on the right side of the tongue. This was surgically excised and histologically diagnosed as a Spindle cell Sarcoma. Results of bone marrow aspiration further revealed that about 16% of his bone marrow was infiltrated with abnormal pleomorphic vacuolated cells with bluish cytoplasm, consistent with rhabdomyosarcoma (RMS) cells. The child was treated with surgery and chemotherapy with good initial response, but then died after one year following a relapse.

Key words: Spindle Cell, Sarcoma, Tongue, Male, Child.

# **INTRODUCTION**

Spindle cell tumour is a rare soft tissue lesion that can be benign or malignant. Malignant lesions can present as carcinomas, sarcomas or carcinosarcomas. Spindle cell sarcoma is an extremely rare soft tissue tumour and only a handful affecting structures in the oral cavity has been reported in the English literature.<sup>1</sup> Whereas in adults the commonest site is in the head and neck region<sup>2</sup> it is mostly found in the paratesticular region in children.<sup>3</sup> The commonest site when it does occur in the head and neck region, include the orbit, nasopharvnx, paranasal sinuses, cheek, neck, middle ear and larynx.<sup>1</sup> Occurrence of this tumour in the tongue, especially in children has very rarely been reported.<sup>3,4</sup> We report here an unusually massive, pendulous Spindle cell sarcoma arising from the tongue in a five-year-old male child that, after resection, weighed 989 grams about 10% of the patient's total body mass.

## Case Report

A 5-year-old male patient presented to the department of child health, Korle Bu Teaching Hospital, with a huge and painless tumour of the tongue of nine months duration. His past medical history was unremarkable. Clinical examination showed a massive lesion of the tongue which hung like a pendulum onto his chest. It was non-tender except at the posterior aspect of its lowermost part which appeared inflamed and covered with a dark discoloured necrotic scab (Figure 1). Physically, the patient appeared very cachectic, (weight 9.9kg) and anaemic. He was unable to feed properly, but managed to swallow liquid diet administered orally with a spoon.



Figure 1 Spindle cell sarcoma of the tongue of nine months duration at the time of presentation

There were palpable, mobile and tender bilateral submandibular lymph nodes and hepatosplenomegaly. The patient was sickle cell negative. His haemoglobin was 6.8g/dl and his red blood cells showed hypochromasia. His differential white cell count was within normal limits. Chest x-ray revealed clear lung fields, but abdominal ultrasound confirmed hepatosplenomegaly. Neither CT scan nor MRI of the lesion was done because these facilities were unavailable at the time.

#### **Biopsy and Diagnosis**

Incision biopsy specimens were taken at two different areas of the lesion under local anaesthesia for histopathological investigations. The pathologist reported "an ulcerated tumour composed of elongated fusiform cells associated with abundant mitotic figures. The stroma made up of prominent collagen fibres, some myxoid areas and prominent vasculature". A diagnosis of spindle cell sarcoma of the tongue was made (Figure 2). Immunohistochemistry was not done due to lack of this facility at our institution; however the results of bone marrow aspiration revealed, "- the patient has about 16% his bone marrow infiltrated with abnormal pleomorphic vacuolated cells with bluish cytoplasm, consistent with Rhabdomyosarcoma cells." The patient was treated as a case of advanced highgrade rhabdomyosarcoma.

#### Surgery and Chemotherapy

Under general anaesthesia with naso-tracheal intubation, after his haemoglobin level was brought up to 10.5 g/dl with blood transfusion, the tumour was resected as widely as practical leaving enough normal tongue tissue to allow for reconstruction of the tongue for function.



**Figure 2** Photomicrograph of spindle sarcoma (H&E x 400) showing elongated fusiform Spindle cells (White arrow) arranged in fascicles with frequent mitosis (Black arrow)

The lip was found deformed at the right commissure due to pressure from the tumour mass (Figure 3). The excised tumour mass measured 19.5cm by 14.0 cm by 12cm and weighed 989 grams, about 10% of the patient's total body mass at the time of surgery.



Figure 3 Photograph showing remarkable initial recovery of patient four months after surgery and chemotherapy

Commencing twelve days after surgery, he received nine cycles of combination chemotherapy, (Cyclophosphamide Vincristine and Actinomycin D), at three weekly intervals. He responded well to treatment, and was then discharged home, weighing 16kg twenty-nine weeks post-surgery (Figure 3). He returned 6 months later to the paediatricians, with a relapse in the tongue and metastatic spread to the submandibular region and cervical lymph nodes. There was no response to further chemotherapy and he died at home soon after.

# DISCUSSION

Soft tissue sarcomas arising in the head and neck are very rare accounting for 10% of all sarcomas and approximately 1% of head and neck tumors.<sup>5</sup> maxillofacial sarcomas constitute between 4-8% of all malignancies in the head and neck region<sup>6</sup>. There is a bimodal distribution of presentation with an initial peak incidence between 2-5 years of age and a second surge at 10-19 years. RMS belongs to the class of "small round blue cell tumour of childhood." It is classified as embryonal, alveolar and pleomorphic.<sup>7</sup> The spindle cell variant was first described in 1992, and is associated with good prognosis.<sup>8</sup>

It is uncommon and is mostly found in the paratesticular region in children.<sup>3</sup> The patient in this case was five years old, and the lesion involved the tongue and was enormous at the time of presentation. The massive size of the tumour, constituting 10% of the patient's total body mass at the time he reported for treatment, is remarkable and far exceeds the dimensions of any of the previously reported cases in the literature.<sup>3,4,9,10</sup>

Clinical findings, diagnostic evaluation and therapy depend upon location of the primary tumour and age of the patient.<sup>7</sup> Current treatment of these lesions is by the use of multi-modal approach in which surgery is combined with either radiotherapy or adjuvant chemotherapy.<sup>11</sup> The prognosis for a child or adolescent with rhabdomyosarcoma is related to the age of the patient, site of origin, widest diameter of the tumour, resectability, presence of metastases, number of metastatic sites or tissues involved, presence or absence of regional lymph node involvement, and histopathological subtype.<sup>12-14</sup> Children with metastatic disease at diagnosis have the poorest prognosis.<sup>15,16</sup> In this case, the patient was clinically unwell and had metastatic disease at presentation. He was managed with surgery and chemotherapy. The child responded very well clinically after the initial phase of treatment.

However, he returned 6 months later, with a relapse in the tongue and metastatic spread to the submandibular region and cervical lymph nodes, with no response to further chemotherapy.

## CONCLUSION

Despite the very late presentation of this tumour as evidenced by the enormity of the size of the tumour, and the distant metastasis to the bone marrow, the therapeutic outcome employing surgery and chemotherapy was remarkable and lead to a rapid improvement of quality of life in the child.

#### Consent

Written informed consent was obtained from the patient's mother for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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