

# Primary alveolar rhabdomyosarcoma: a horrendous presentation with miraculous remission

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## DESCRIPTION

Alveolar rhabdomyosarcoma constitutes approximately 1% of all paediatric tumours. Primary alveolar rhabdomyosarcoma of lip is a very rare pathological entity.<sup>1</sup> Here is presented a case of primary alveolar rhabdomyosarcoma of upper lip treated with palliative external radiation therapy followed by combination chemotherapy.

A 5-year-old girl presented with painful swelling of upper lip of 3 months duration, which progressed rapidly. There was no history of other swelling. There was no history of any malignancy in the family. Local examination revealed an ulceroproliferative growth of 10×8 cm size present over the upper lip involving bilateral nostrils and was locally advanced, causing difficulty in swallowing and respiration (figures 1 and 2). Histopathological examination confirmed the diagnosis of alveolar rhabdomyosarcoma.

The poor general condition of the patient as well as obstructive symptoms prompted us to treat the patient with palliative intention. The patient received palliative external radiation therapy and showed partial regression of the swelling after radiotherapy. The patient was further planned for intravenous combination chemotherapy with vincristine 1 mg, actinomycin 0.5 mg and cyclophosphamide 400 mg 3 weekly for six cycles and after



**Figure 2** Lateral view of the growth.

6 months of the initial presentation, the patient was in complete remission as is evident from the serial photographs (figures 3–5). Malignancy at this rare site is uncommon and clinical management presents considerable challenges.<sup>1</sup> Intensive multimodality approach should be tested, incorporating surgery, dose-intensive combination chemotherapy and radiation therapy.<sup>2</sup> Future challenges include the



**Figure 1** Anterior view showing a locally advanced growth over the upper lip involving bilateral nostrils.



**Figure 3** Clinical photograph at the end of palliative radiotherapy.

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**Figure 4** Post-treatment photograph after 2 months of completion of radiotherapy.

development of less toxic therapy for patient with localised disease and new approaches for patients with metastatic disease.

### Learning points

- ▶ Primary alveolar rhabdomyosarcoma of the lip is a rare pathological entity and usually present in the aggressive form.
- ▶ External radiotherapy and adjuvant combination chemotherapy with vincristine, actinomycin and cyclophosphamide is very effective treatment protocol.
- ▶ Intensive multi-modality approach for treatment is recommended as the results are miraculous and complete remission of the disease is possible.



**Figure 5** Post-treatment photograph showing complete remission after 6 months of completion of radiotherapy and six cycles of chemotherapy.

**Competing interests** None.

**Patient consent** Obtained.

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