

REPORT OF A GIRL WITH INFANTILE HEMANGIOENDOTHELIOMA

Köksal Yuca¹, Mustafa Kösem², Sevil Arı Yuca³, Hayal Oral², Ali Bay⁴

Yüzüncü Yıl University Faculty of Medicine, Departments of Otorhinolaryngology¹, Pathology² and Pediatric Hematology⁴, Özalp Training Hospital, Clinic of Pediatric Diseases³, Van, Turkey

Infantile Hemangioendothelioma (IHE) is a rare vascular tumor with predominant involvement of soft tissues of extremities. The occurrence in the head and neck region is extremely rare. We report a four-month-old female with IHE in her lower lip. The clinical and histologic photographs depict the findings. The diagnosis and treatment of IHE are discussed.

Key words: Hemangioendothelioma, vascular tumor, infants.

Eur J Gen Med 2005; 2(2):80-82

INTRODUCTION

Infantile hemangioendothelioma (IHE) is a severe disease with a high mortality. It is characterized by multiple hemangiomas affecting the skin and visceral organs (1). Hemangioendothelioma is a rare vascular tumor of endothelial cell origin show an intermediate malignant potential between simple hemangioma and angiosarcoma. The tumor is characterized by neoplastic proliferation of epithelioid or histiocytic endothelial cells. It may involve bone or soft tissues of extremities and can behave like a benign or malignant tumor. The occurrence in the head and neck region is a rare and believed to be a neoplasm of borderline malignancy. In the literature, there are several case reports on the involvement of the head and neck region (2-5). For primary treatment recommendation there is general agreement in the literature as surgical excision of the tumor (2-4, 6).

We reported a case of a 4-month old infant with IHE resulted in soft tissue destruction of lower lip and histologic findings were described.

CASE

A 4-month old girl with a lower lip mass since birth which began to grow two months ago was admitted to our clinic. Clinical examination showed an ulcerated lesion of 5x2 cm in size in her lower lip, starting from the

right mouth commissura, extending through the midline and vermillion and including a part of the gingiva (Figure 1).

No palpable lymphadenopathy was observed in the neck. Outer ear, nose, and throat examination, general physical examination, and routine laboratory investigations showed no abnormality. A punch biopsy was performed under local anesthesia. Histologic examination revealed IHE; the tumor consisted of plump endothelial cells, stromal cells, and residual striated muscle tissue. The surface epithelium was ulcerated. Mitotic figures were scant (Figure 2). Surgical resection was recommended but refused by the patient's family. The child did not return for any further treatment.

DISCUSSION

Hemangioendotheliomas are rare vascular tumors show an intermediate malignant potential between simple hemangioma and angiosarcoma (2). The tumor is characterized by neoplastic proliferation of epithelioid or histiocytic endothelial cells. Predominant locations are soft tissues of extremities, the liver and lungs; involvement of the head and neck region is a rare (2). The biologic behavior of the tumor appears to be dependent on its location. Mortality from epithelioid hemangioendothelioma in the

Correspondence: Köksal Yuca, MD,
Yüzüncü Yıl University, Faculty of Medicine, Department
of Otorhinolaryngology, 65200, Van-Turkey
Tel: 904322157168, Fax: 904322166563
E-mail: koksalyuca@yahoo.com



Figure 1. The ulcerated lesion of 5x2 cm in size in the lower lip.

soft tissues was 13%. As of January 2000 a total of 17 cases with involvement of the head and neck region have been described in the literature; only six case reports with intraoral hemangioendothelioma in the pediatric population have been published(2). Our case had an ulcerated lesion of 5x2 cm in size in her lower lip, starting from the right mouth commissure, extending through the midline and vermilion and including a part of the gingiva. Outer ear, nose, and throat examination, general physical examination, and routine laboratory investigations showed no abnormality.

Hemangioendothelioma is a confusing term. It has been used over the years for vascular tumors composed of endothelial cells, which may involve bone or soft tissues (3, 5, 7). In 1908 Mallory introduced the term hemangioendothelioma (3, 7). Since then, various other synonyms have been adopted including, infantile hemangioendothelioma. Types 1 and 2, cellular angioma of infancy, benign hemangioendothelioma, non malignant hemangioendothelioma, angioendothelioma, lymphangiosarcoma, hemangiosarcoma, angiosarcoma, angioblastoma and intravascular endothelioma (2, 3, 7).

Bones of the limbs are most commonly involved. We are aware however, of case reports on the involvement of the head and neck region, as well (3, 5, 7, 8). Our case with a lower lip mass since birth that began to grow at the age of two months was clearly unusual.

A definitive diagnosis can be made by means of a biopsy only. Histopathologically hemangioendothelioma is composed of

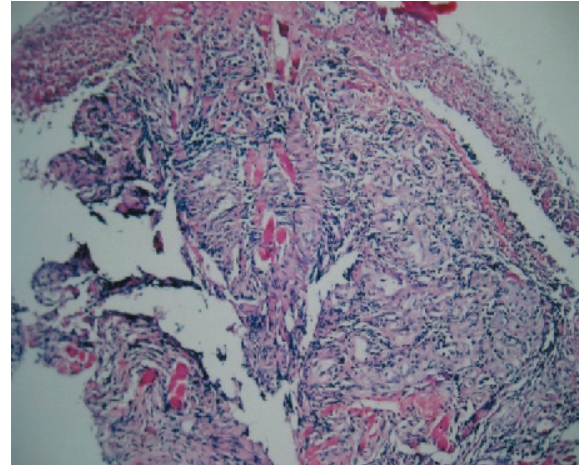


Figure 2. Tumor tissue consisted of well-formed blood vessels, proliferating endothelial cells, residual striated muscle tissue, and necrotic exudates on the surface (H-E X 125).

short strands or solid nests of rounded to slightly spindled endothelial cells, which form small inconspicuous lumina, containing erythrocytes. In most cases this tumor shows only mild nuclear pleomorphism and virtually no necrosis or mitotic activity (3).

Treatment of hemangioendothelioma usually consist of wide radical surgical excision (2-4,6,8,) with postoperative radiotherapy(3,7), especially in high- grade lesions. Radiation therapy has been used alone when surgery was not feasible (7). Chemotherapy currently has no significant role in the treatment. Orchard et al reported successful treatment with α -interferon in two infants with hemangioendotheliomas, one in the head and neck region (7). In our case; surgical resection was recommended but refused by the patient's family.

In summary due to the confusing nosology with unpredictable biologic behavior and high mortality rate IHE requires close follow-up and urgent surgical treatment of the patient.

REFERENCES

1. Park EA, Seo JW, Lee SW, Choi HY, Lee SJ. Infantile hemangioendothelioma treated with high dose methylprednisolone pulse therapy. *J Korean Med Sci* 2001;16: 127-9
2. Eckardt A, Kuettner C, Kuske M. Hemangioendothelioma of mandible in a newborn infant-diagnostic and treatment approach of a rare vascular tumor. *Oral Oncol* 2001; 37:668-72

3. Eliashar R, Saah D, Osin P, Sichel J. Hemangioendothelioma of the temporal bone in a child. *Int J Pediatr Otorhinolaryngol* 1997;40:67-71
4. Hamakawa H, Omori T, Sumida T, Tanioka H. Intraosseous epithelioid hemangioendothelioma of the mandible: a case report with an immunohistochemical study. *J Oral Pathol Med* 1999;28:233-7
5. Cansiz H, Yener M, Dervisoglu S, Kalekoglu N. Hemangioendothelioma of the frontal bone in a child. *Craniofac Surg* 2003;14:724-8
6. Bay A, Öner AF, Etlik Ö, Koseoglu B, Kaya A. Unusual Presentation of Infantile Hemangioendothelioma. *Pediatr blood and cancer*. 2005 impress
7. Ibarra RA, Kesava P, Hallett KK, Bogaev C. Hemangioendothelioma of the temporal bone with radiologic findings resembling hemangioma. *Am J Neuroradiol* 2001;22:755-8
8. Carlotti CG Jr, Jay V, Rutka JT. Infantile hemangioendothelioma of the pericranium presenting as an occipital mass lesion. Case report. *J Neurosurg* 2000;92:156-60