REPORT OF A GIRL WITH INFANTILE HEMANGIOENDOTHELIOMA

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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.

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 showing 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 may behave like a benign or malignant tumor. The occurrence in the head and neck region is 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 resulting 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 vermilion 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 excision was recommended but refused by the patient’s family. The child did not return for any further treatment.

DISCUSSION

Hemangioendotheliomas are rare vascular tumors showing 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 rare (2). The biologic behavior of the tumor appears to be dependent on its location. Mortality from epithelioid hemangioendothelioma in the
Infantile hemangioendothelioma in a girl.

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 commissura, 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 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