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Case Report

LOBULAR CAPILLARY HAEMANGIOMA OF THE NASAL CAVITY IN CHILDREN

Literature survey and case report

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ABSTRACT

A case of a child with lobular capillary haemangioma (LCH) in the nasal cavity along with a concise literature survey is presented. This congenital lesion is typical of the skin and oral mucosa and uncommon for paranasal sinuses and nasal cavity. They represent a common reactive angiomatous proliferation that occurs in the skin and mucous membranes. The pathogenic endothelial cells in infantile haemangioma are of unknown origin. Haemangiomas are classified as capillary, cavernous, and mixed lesions. The treatment remains an area of considerable controversy. New therapeutic options include laser therapy, systemic and intralesional corticosteroids, interferon, sclerotherapy, cryotherapy and improved surgical interventions.

Key words: lobular capillary haemangioma, nasal cavity, diagnosis, treatment.

INTRODUCTION

Haemangiomas are benign tumours that originate in the vascular tissues of skin, mucosa, bone, muscles and glands (1) Traditionally, this term has been applied to benign tumours of vascular tissues and vascular malformations. In the biologic classification proposed by Mulliken & Glowacki in 1982, haemangiomas are defined as vascular tumours that enlarge by rapid cellular proliferation. They are classified as capillary, cavernous, and mixed lesions. Usually, congenital lesions are located on the skin or oral mucosa while the nasal cavity and paranasal sinuses are uncommon sites for haemangiomas. Nasal cavity haemangioma is extraordinarily rare in children (2), (3), (4). Only nine cases have been reported in the English literature between 1985 and 2005 (5).

We report a child with lobular capillary haemangioma (LCH) of the nasal cavity, its surgical treatment and add a literature survey on LCH as well.

CASE REPORT

An infant of male gender and aged 1 year and 9 months who was hospitalized in the Clinic of Otorhinolaryngology, St. Marina University Hospital of Varna following frequent epistaxis, hampered breathing and nasal apex deformity was reported. The inspection of the external nose revealed swelling of the nasal apex, while the anterior rhinoscopy detected a tumour of nodular shape originating from the margin of Mink's valve and considerable nasal cavity obstruction. It was of pink-red colour, abundantly blood-supplied and mobile on palpation. CT examination showed that the neoplasm originated from the area of nasal apex and freely entered the nasal cavity. Surgical intervention was performed under general anaesthesia through an open access. The neoplasm originated from the caudal edge of triangular cartilages and from the cephalic edge of the alar cartilages and from the upper margin of the nasal septum. During the operation, the bleeding was not intensive and thus the neoplasm was radically removed. The histological examination of the specimen demonstrated that the tumour was formed by vessels of capillary type located among the hypercellular stroma. The capillary formations were covered by one-two endothelial cell layers situated on a basal membrane. The

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vascular lumens were smaller or larger in size. Some areas of cavernous structure were observed on the periphery. LCH was concluded on histological basis. The postoperative period was smooth and the one-year long following-up did not indicate any relapse of the disease at all.

INCIDENCE RATE AND LOCALIZATION

Between one-third (6), and up to one half (7) of the cases are present at birth while the rest ones occur early in infancy. Sixty per cent of haemangiomas resolve by 4 years and 76 % do by 7 years. In newborns, the incidence rate is 1-3% and increases up to 10% by the age of 1 year (8). Whereas 80% of the patients present with a single haemangioma, others have multifocal ones. Infants with multifocal lesions are at a higher risk of having visceral haemangiomas, especially intrahepatic ones (9). The nasal cavity and paranasal sinuses are extremely rare sites for this tumour in children (3).

Aetiology

Recent studies with isolated haemangioma (10) and whole lesions (11) indicate that the neoplasm arises from uncontrolled clonal expansion of the endothelial cells. The origin of the pathogenic endothelial cells in common infantile haemangioma is unknown. (12). The transcriptomes of human placenta and infantile haemangioma are sufficiently similar to suggest a placental origin for this tumour, expanding on recent immunophenotypical studies having suggested this possibility (13). In a series of 40 patients with LCH, pregnancy was identified as possible cause in two cases (5%) (14).

Clinical symptoms

Haemangiomas are clinically heterogeneous, their appearance is dictated by the depth, location, and stage of evolution as well as by patient's age. Although rarely life threatening, haemangiomas can pose serious concerns to the cosmetic and psychosocial development of the afflicted child. Visceral haemangiomas carry a much higher morbidity and mortality rate (40–80%), as high flow lesions, such as those found in the liver, may induce high output cardiac failure and anaemia (7).

In the nasal cavity, the haemangioma appears as a node (2), (5) up to 3 cm in size (29). It can be of infiltrative growth not only towards the septum, but also to the lateral side of the nose, even in children (16). Lesions vary in size from 1 to 8 cm and mainly involve the nasal septum (in 45%) and the nasal vestibule (in 17,5% of the cases) (14). The symptoms consist in difficult nasal breathing, epistaxis, epiphora, purulent or mucous rhinorrhoea and cosmetic alterations of the external nose. Unilateral epistaxis occurs in 95% while nasal obstruction - in 35% of the patients with LCH (14).

Complications of haemangiomas are cosmetic and functional. They depend on the location, size, or rapid proliferating phase of the neoplasm. Some type of complication is found in 40% of lesions, the commonest being ulceration (21%) and bleeding (7,5%) (15).

Histology

Haemangiomas are heterogeneous at all stages of tumour development (16). LCH was histologically characterized by lobular proliferation of capillary blood vessels (17), (18). Proliferating lesions consist of endothelial cells (EC), supporting pericytes, and myeloid cells. They include other cells such as fibroblasts and mast cells, too (16).

Differential diagnosis

These lesions may be confused with a variety of other benign vascular tumours such as angiofibroma, angiofibromatous polyp, leiomyoma, and glomus tumour, as well as malignant fibrous histiocytoma and angiosarcoma, haemangioendothelioma (19). Juvenile haemangioma (a cellular haemangioma of infancy) is a histologically similar lesion to LCH, although its clinical features are thoroughly different (20).

Treatment

The management of haemangiomas continues to be an area of considerable controversy (7). The treatment of congenital vascular anomalies is based on an understanding of the clinical behaviour and natural history of individual lesions (21), the localization and size of the neoplasm, the stage of development as well as patient's age.

1. *Intranasal injections of sclerosing solutions*

   Sclerotherapy is effective in more than 90% of haemangiomas, but not in involuting lesions. Superficial lip and eyelid lesions and bulky haemangiomas of subcutaneous tissue are treated with sclerotherapy without complications other than very minimal epithelial desquamation (22).

2. *Intralesional injections of steroids*

   Intralesional steroid injection is used during the proliferative phase of tumour growth. They may be appropriate and effective for small, localized cutaneous haemangiomas (23). The complications include sclerodermiform atrophy, eyelid necrosis, and central retinal artery occlusion (23), skin necrosis and adrenal suppression (24), (25), as well as local ulceration of the lesion (26).

3. *Subcutaneous interferon application*

   The more recent options of interferon-alpha (IFN-alpha) 2-a or interferon alpha 2-b are effective, however, about 20% of the infants develop spastic diplegia (4). Its potential side effects include fever, malaise, leukopaenia, interstitial nephritis and haemolytic anaemia (27).

4. *Systemic therapy with steroids and IFN-alpha*

   Systemic therapy with steroids or INF-alpha is successfully administered in infants with cutaneo-visceral haemangiomatosis presenting with life-threatening complications (21). Oral prednisolone is given in the dosage of 5 mg/kg/day for 6-9 weeks, then 2-3 mg/kg/day for 4 weeks, and followed by alternate day therapy for up to 6 weeks. Treatment with systemic steroids in infants has a risk of growth suppression, diabetes mellitus, hypertension and rebound haemangioma growth after treatment cessation (28).

5. *Laser therapy*

   Several laser systems have been used to treat intact and ulcerated haemangiomas. Optimal laser delivery systems and parameters selective for treatment of haemangiomas have yet to be defined (7).

6. *Cryosurgery and x-ray therapy*

   Cryotherapy is popular in certain parts of Europe and South America and produces favourable results with superficial haemangiomas. However, concern about potential scarring limits its use in North America (7). The use of radiation in the management of haemangiomas is controversial owing to the high complication rate (29).

7. *Cytostatics (chemotherapeutic means)*

   When a life-threatening haemangioma is resistant to corticosteroids, vincristine may be an option. There are several reports of benefit in large, life- or function-endangering haemangiomas (30), (31).

8. *Embolization*

   Embolization is effective for large lesions that are not amenable to other treatment options or in preparation for surgical excision (32).

9. *Surgical treatment*

   After a thorough physical examination, investigations may be indicated either if complications are present, or if surgical intervention is contemplated (21). Surgical excision is used most frequently later in childhood to correct residual scarring or to remove fibro-fatty tissue. Early excisional surgery is, however, a reasonable option in selected cases such as pedunculated haemangiomas where residual abnormality is virtually inevitable and in function- or life-threatening haemangiomas where pharmacologic therapy is not effective or well-tolerated.

The present authors recommend re-evaluation at about 4 years of age to assess how much residual haemangioma is present and consider surgery if it is likely to give a good result in a haemangioma causing scarring or involuting very slowly (7). A surgical approach to the haemangioma may be indicated in an attempt to gain a more normal appearance.

Most haemangiomas do not require immediate intervention and 90% can be expected to undergo gradual involution before the age of 9 years (33). Approximately 20-40% of the patients will have residual and disfiguring skin changes (7). In 50% of these cases, normal skin is restored, whereas in the remaining ones, the cosmetic result is much better than that offered by surgical intervention (8). Surgical removal of haemangiomas is recommended when the tumour causes functional and aesthetic discomfort. The transcolumellar incision in rhinoplasty has proven to be a safe and effective technique (34). In this particular case the transcolumellar incision is performed in order to obtain full access to the nasal dome and safe removal of the tumour.

CONCLUSION

Corticosteroids and IFN form the first line of treatment (21). Vincristine and bleomycin are considered for problematic haemangiomas in infants failing to respond to steroids. Surgery is indicated in small, well-localized lesions of the eyelid, lip, and neck or other parts of the body and when it is likely to lead to an aesthetically more acceptable scar.

In our opinion, the choice of treatment of childhood haemangiomas should be considered for each individual case. It depends on patient's age, localization, stage of disease, risk of relapse, formation of visible scars and functional and life-threatening conditions as well.

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