A 22-month-old boy with painless swelling of the left side of face

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Article Info

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Received: 25 January 2020
Accepted: 18 February 2020
Available Online: 19 February 2020

ISSN: 2224-7750 (Online)
2074-2908 (Print)

DOI: 10.3329/bsmmuj.v13i1.45034

Keywords: Face; Lymphangioma; Pain: Swelling

Cite this article:
Akhter M, Imon AA, Hossain R, Sobhan SM, Haider G. A 22-month-old boy with painless swelling of the left side of face. Bangabandhu Sheikh Mujib Med Univ J. 2020; 13: 27-30.

Available at:
www.banglajol.info

A Journal of Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

Presentation of Case

Dr. Mahmuda Akhter (Associate Professor): A 22-month-old boy came to the outpatient Department of Oral and Maxillofacial Surgery at Bangabandhu Sheikh Mujib Medical University with the complaints of painless swelling on the left side of the face since birth (Figure 1). Initially, it was small but gradually increased in size. On examination, there was a soft, non-tender, non-pulsatile swelling on the left side of the face causing facial asymmetry, causing the nose and upper lip to deviate to the opposite side. It extended from the left-sided upper eyelid to the lower border of the mandible of the same side, which is around 7 x 4 cm in size with diffuse border. Temperature, color and texture of the skin over the swelling were normal. There was a gross paraxial proptosis of the left eye. The eyeball level was different. The vision was normal by the pediatric assessment. On intra-oral examinations, the mucosa was normal in color and texture. Dentition and occlusion were normal. Mouth opening was normal. The patient had no difficulties in eating, swallowing or speech. Lymphadeno-pathy was absent with no other systemic abnormalities. No suggestive family history was found.

Ultrasonography of the swelling reveals large diffuse mixed echogenic soft tissue mass having multiple intercommunicating tubular structures seen in left cheek. No vascular flow is identified. Also, in color doppler ultrasonograph of the lesion reveals soft tissue mass extends into the left upper eyelid and upper lip. The average thickness of this area is about 14 mm. There is the presence of lobulated dilated channels seen within this mass. The diameter of the dilated channels ranged from low as 2.7 mm to highest around 7.6 mm. with no flow within channels, compatible with lymphangioma.

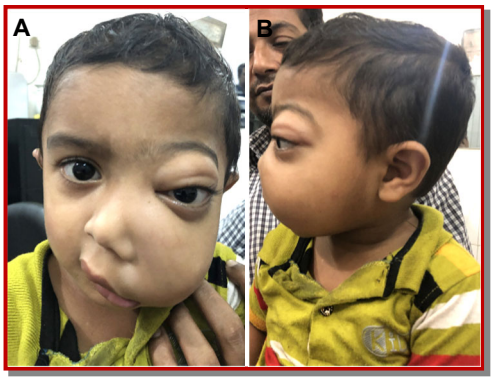

Figure 1: Painless swelling on the left side of the face since birth. Front view (A) and lateral view (B)

In magnetic resonance imaging, there was a large thin septate T1 hypo, T2 fat sat hyper-intensity lesion (7 x 3 cm) seen in the left side of the facial subcutaneous lesion, extending to pre-septal to pre-maxillary area (Figure 2). Few linear (1 cm) lesions seen in left retrobulbar intraconal spaces with proptosis. With contrast magnetic resonance imaging, significant enhancement of the lesion seen which is compatible with subcutaneous lymphangioma.

On fine needle aspiration cytology, it shows many lymphocytes in the soft tissue with proteins materials which match with lymphangioma.

With the co-relations between clinical, imaging and histopathological findings, diagnosis of lymphangioma of the left-sided facial region was considered.

Provisional Diagnosis

Lymphangioma of the left cheek

Differential Diagnosis

Lymphangioma

Lymphangioma is a congenital malformation of the lymphatic vessels common in the head neck region. They consist of a center of abnormal development of the lymphatic tissue.[1] This is a benign type of vascular malformation. Lymphangiomas are unencapsulated and histologically composed of fibrous materials with endothelial lacunae usually filled with blood or serious fluid,[2] thereby they present as a soft, compressible, lobulated ill-defined mass which is not attached to the skin. The incidence of lymphangioma is 1.2–2.8/1,000 newborn babies.[3] The clinical presentation of this patient is relevant to all these features.

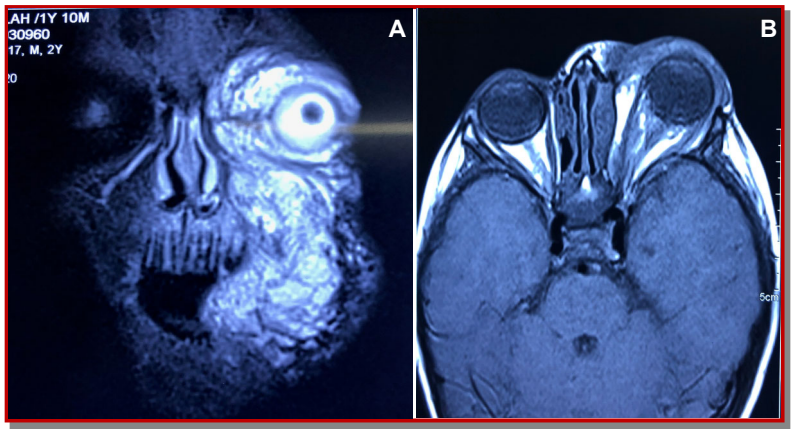

Figure 2: Magnetic resonance imaging coronal view (A) and axial view (B)

Lymphangioma circumscriptum

Dr. Ashik Abdullah Imon (Assistant Professor): This is a benign type of microcystic lymphatic malformation. The pattern of this lesion consists of small blisters which are usually either pink or dark red.[4] Treatment is not required except aesthetic reason.

Cavernous hemangioma

Prof. Golam Haider: Cavernous hemangioma is a congenital condition and increases in size with the ages. cavernous hemangiomas consist of large, dilated vascular channels in diffuse patterns. They often show thrombosis, perivascular hemosiderin deposition and calcifications.[5] In this condition, blood flow is slow down and intercellular junctions cause leakage of fluid. Cavernous heman-giomas do not tend to regress. Magnetic resonance imaging is recommended to confirm the diagnosis. Lymphan-gioma differentiated from cavernous hemangioma by the presence of lymphoid aggregation in the stroma with irregular lumens, widely spaced nuclei whereas in cavernous hemangioma, large dilated blood-filled vessels lined by flattened endothelial cells. Vessels are arranged roughly in a lobular or diffuse haphazard pattern.

A-V malformations

Dr. Ryfat Hossain (Assistant Professor): These are shunts like anastomoses due to disorders in angio-genesis.[6] This malformation develops due to direct communications between arteries and veins by-passing capillary anastomoses. So, there is a disruption of the natural blood flow. The vulnera-bility of this malformation depends on the size and number of blood vessels involved. On palpation, it is warm, pulsatile and painful, which may be due to frequent bleeding. CTA, MRA, catheter angiogram needed to confirm the diagnosis. For this patient, there was no history of bleeding, raised local temperature and sign of pulsation.

Orbit temporal neurofibromatosis

Dr. Sheikh Mahbub-Us Sobhan (Associate Professor): Neurofibromatosis in orbit, temporal and facial region known as orbit-temporal neurofibromatosis.[7] Children and adults with neurofibromatosis type 1 (NF1); an autosomal dominant condition, present some ophthalmologic signs and symptoms like pedunculated upper eyelid. Gross dimness of vision with or without proptosis. Plexiform neurofibromas (PNs) involving the eyelid, orbit, periorbital, and facial structures cause visual loss in children secondary to proptosis, ptosis, and facial disfigure-ment. MRI excludes the diagnosis.

Dr. Akhter's Diagnosis

Lymphangioma

Discussion

Dr. Imon: The incidence of lymphangiomas ranges from 1.2 to 2.8 per 1000 newborns.[3] Lymphangioma appears at birth or early in life. About 50% of lymphangiomas are seen at birth, and most lymphangiomas are evident by the time the patient is aged 5 years. It also documented in fetuses too.[8] Though Lymphangiomas are frequently seen in the head and neck region but can occur throughout the body particularly in the proximal extremities, buttocks and trunk. These develop in early embryonic life when the lymphatic vessels do not form properly.

Usually, lymphangioma are two types: cavernous lymphangiomas and cystic hygromas. Cavernous lymphangiomas are found in areas, such as the tongue and floor of the mouth. Cystic hygromas, on the other hand, arise from lymphatic tissue in areas where expansion can occur and large multiloculated cystic spaces can develop. Based on cystic space size, they are classified as: macrocytic, microcystic and mixed.[9] Histological features are dilated lymphatic channels with one or two endothelial layers, with or without an adventitial layer.

Dr. Masud Bin Hasan (Resident student): How we can diagnose lymphangioma?

Dr. Akhter: Diagnosis is based on clinical examina-tion, imaging and histopathological evaluation. These lesions present as a mass that is soft compressible, loculated and ill-defined. The lesion is free from the skin or movable across deeper tissues and readily transilluminate.[10] The ultrasono-gram, computer tomography and magnetic reso-nance imaging use to define the relationship of the lesion with the surrounding structures and to help plan surgical strategies.[10] We also go for fine needle aspiration cytology to confirm the histological diagnosis of the disease. Lymphangiomas can spon-taneously regress or may become aggressively invasive lesions as is apparent in our case. Among

the complications, spontaneous or traumatic hemorrhage may occur.[11] In adult patients, neoplasm can switch to squamous cell carcinoma.[12]

Dr. Md. Wahidujjaman (Assistant Professor): Why you give priority on treating ocular conditions?

Dr. Sheikh Mahbub-Us Sobhan (Associate Professor): Because the lesion causes proptosis, ptosis of the eyelid, restricted ocular movement and compression of the optic nerve leading to enormous visual disabilities. A study showed 26 patients with orbital lymphangiomas, 85% had proptosis, 73% ptosis and 46% had restricted movement.[13]

Dr. Shakhawat Hossain Sayantha (Assistant Professor): Why do you prefer bleomycin and what are the other treatment option?

Prof. Haider: Despite the presence of multiple treatment modalities, adequate excision of the lesion can be quite challenging due to its infiltrative nature and so local recurrences are common. Other options like cryotherapy, sclerotherapy, and cautery and radiofrequency therapy have also been reported with different degrees of success.[14] Bleomycin acts as a sclerosing agent and causes fibrotic transformation of vascular endothelium. Some article reveals that 90% regression occurs in 53.4% and more than 50% regression in 26.7% case after application of bleomycin.[15] With limited complications, intralesional bleomycin injections have proven to be quite effective in similar cases.[15, 16] Adverse effects like fever, vomiting[16] are usually manageable.

Dr. Sarwar Jamal Biplob (Resident student): When you will start the treatment of cheek conditions?

Dr. Hossain: Lymphatic malformation may regrow in some cases. Mainly to avoid recurrence along with the extreme age (22 months), and post-traumatic syndrome, we decided to wait for one to two years.

Figure 3: After first dose of bleomycin intralesional injection (A); After second dose of bleomycin intralesional injection (B); 3 months after surgery (C)

Final Diagnosis

Lymphangioma

Treatment

The treatment was as follows: a) Treatment for the eye to salvaging the visual outcome (Phase 1); b) To observe one to two years to assess the improvement of anatomical and functional eye condition and recurrences, followed by treatment for cheek (Phase 2).

Prof. Haider: Phase 1 treatment: Intralesional injection of bleomycin was selected as a treatment of choice as a sclerosing agent- Injection bleomycin 0.5 mg/kg intralesionally under general anesthesia after aspirating lymphatic fluid as much as possible by 10 mL syringe in the ratio of 5:1 (aspirated fluid: Injection bleomycin). The vial contained 15 units of bleomycin. The injecting dose did not exceed 10 units at a single dose. The first dose was given and advised ultrasonography of the lesion with color doppler after 4 weeks. In a follow-up after four weeks, the improvement was satisfactory, so the second dose was given. But after a few days, the condition worsened. It was decided to go for the surgery. With the upper eyelid incision, surgical removal of the growth was done, though complete removal was not possible. The boy's parent was advised to follow-up as per protocol (Figure 3).

Follow-up is continued every six months.

Conflict of Interest

Authors declare no conflict of interest.

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