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Case Report

Facial Malformation

Paediatric Facial Malformation - A Case Report

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Abstract: Arteriovenous malformations (AVMs) are rare congenital vascular malformations. They account for 1.5% of all vascular anomalies. Oral and maxillofacial regions are the common sites and account for 50% of the cases. We report a seven-year-old female child with progressive left cheek swelling for the past two years. There was no pain, dysphagia or difficulty in chewing food. But the swelling caused significant cosmetic concern due to facial disfigurement. There was a history of cystic hygroma, operated at a young age. There is no significant family history of vascular anomalies. Child was investigated and diagnosed to have slow flow vascular malformation. Treatment is a multidisciplinary approach. Minimally invasive procedures can conservatively manage it by injecting drugs. Other options are laser therapy, open surgery and a combination of the above treatments. Our case improved after multiple cycles of foam sclerotherapy. Many cases might not improve with one or two injections, but it requires multiple doses. Arteriovenous malformation is not only a physical problem but also involves psychological concern for the child. Though it may be a benign condition, it might cause facial disfigurement, as in our case. A high index of suspicion is required as it is incidentally diagnosed during dental procedures. Multimodality therapy is necessary for the management and is beneficial in all age groups. The present case is published for its rarity.

Keywords: arteriovenous malformation, paediatric, face, sclerotherapy, surgery

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1. INTRODUCTION

Arteriovenous malformations are abnormal dilatations of arteries and veins. These are tortuous interconnections between supplying arteries and draining veins [1]. Arteriovenous malformations (AVMs) are rare congenital vascular malformations. They account for 1.5% of all vascular anomalies. Oral and maxillofacial regions are the common sites and account for 50% of the cases. It might be a cosmetic concern as it can cause facial disfigurement. It can occur anywhere in the body and also in the brain. In brain AVMs, the blood travels directly from the arteries to the veins instead of moving through the capillaries first. This can cause several problems in the brain, including stroke, abnormal fluid balance, hydrocephalus, or, in rare cases, heart failure. Brain arteriovenous malformations (AVMs) sometimes cause no symptoms and are only found during imaging for other reasons. More commonly, though, AVMs can cause bleeding in the brain, seizures, and other neurological problems like persistent headaches, weakness or paralysis on one side of the body, oral bleeding, and problems with speaking. Even though AVMs are rare in children, 3% of them tend to rupture more frequently in children than in adults [2,3]. Most paediatric hemorrhagic strokes are caused due to underlying brain arteriovenous malformations [4]. Multiple risk factors lead to haemorrhage, including previous rupture, big size, deep location of the malformation, deep venous drainage and associated aneurysms [5]. Computed tomography is preferred in ruptured AVMs to find the hematoma's location and size. Magnetic resonance imaging with magnetic resonance angiography is done for better AVM localization and a plan for treatment. Conventional cerebral angiography is the gold standard for diagnosing AVMs as it can show small lesions that are missed in other modalities[6]. Early post-rupture angiography might miss some important components of the AVM due to the compression caused by the hematoma [7,8]. Treatment is a multidisciplinary approach and includes physician, general surgeon, plastic surgeon, radiologist and neurologist. It can be conservatively managed by drugs. Minimally invasive procedures can be done by interventional radiologists. Laser therapy, open surgery and a combination of above therapies is often required.

2. CASE REPORT

A 7-year-old female child, firstborn of a non-consanguineous marriage, was brought to the outpatient with complaints of painless swelling over the left side of her cheek for the past 2 years.

2.1 Medical History

The left cheek swelling was insidious in onset and progressive in nature and attained the present size. History is negative for fever, dysphagia, dyspnea and dental caries. Child was operated for cystic hygroma at one and half years of age. Child is developmentally normal and immunized upto the age.

2.2 Observation

On examination, the child is well nourished. Diffuse swelling of size 10cm X10cm, non-tender, not warm, soft in consistency, non-fluctuant, noticed over the left side of the cheek (Fig.1). It is associated with deviation of the mouth to the right side. The dental examination was normal.

2.3 Family History

No significant family history of vascular malformations

2.4 Investigations

Child was investigated for the same. Blood investigations were normal. MRI Scan of face showed an ill-defined large hyper intense lesion with predominantly T2 hyperintensity with multiple serpiginous areas within the left cheek region in the subcutaneous plane and over the left masseter muscle and minimally extending into the submandibular space suggestive of slow flow vascular malformation (fig.2).

2.5 Diagnosis

Slow flow vascular malformation of the face

2.6 Treatment

A plastic surgeon's opinion was obtained and advised foam sclerotherapy for the same. The child was admitted and eight cycles of sclerotherapy (sodium tetradecyl sulphate) were given as monthly injections.

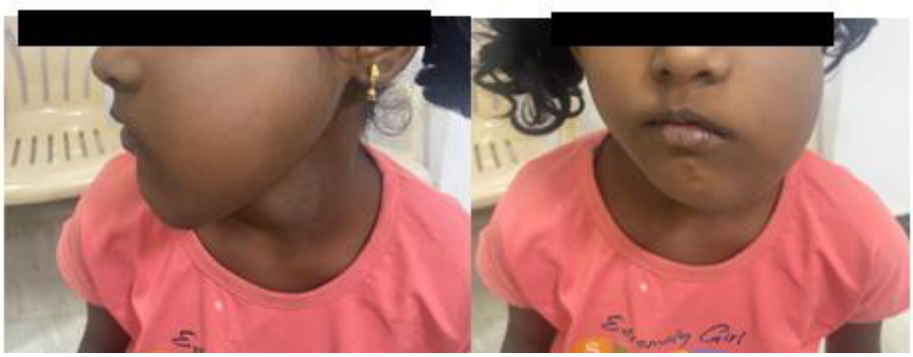

Fig 1: Swelling over the left side of the cheek

Fig.1 Seven-year-old female child presented with diffuse swelling over the left side of the cheek, causing facial disfigurement of size 10cm X10cm, non- tender, not warm, soft in consistency, non-fluctuant, noticed over the left side of the cheek. It is associated with deviation of the mouth to the right side. The dental examination was normal.

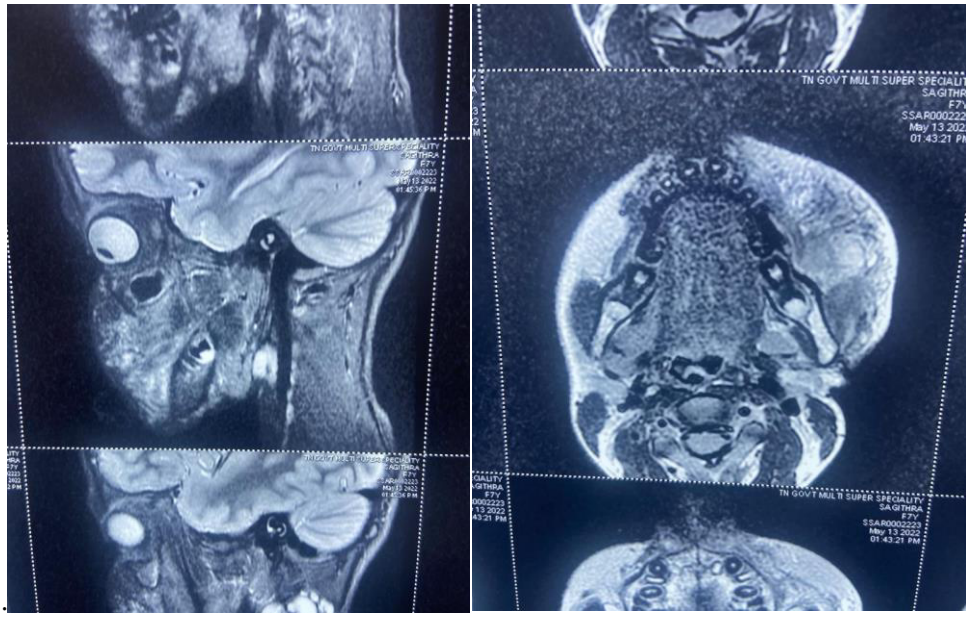

Fig 2: MRI

T2 weighted shows defined large hyper-intense lesion with multiple serpiginous areas within left cheek region in subcutaneous plane and over the left masseter muscle and minimally extending in to the sub-mandibular space. Features suggestive of slow flow malformation.

2.7 *Follow Up*

At present, swelling has regressed and the child is well on follow up. Fig.3

Fig 3: Shows regression of the left side of the cheek swelling post foam sclerotherapy

3. DISCUSSION

Vascular malformations are divided into slow-flow, fast-flow, and complex-combined. Arteriovenous malformations come under the fast-flow category [9]. The most common sites of head and neck arteriovenous malformations are the cheek, ear, and nose. They usually present as bleeding due to trauma or manipulation and there is a high chance of rebleeding[10]. Haemangiomas are classified as infantile or congenital; the Infantile subtype is the most common. Infantile haemangiomas develop during infancy as a subtle punctate lesion. The majority of them grow after birth. Congenital haemangiomas, as the name implies, are fully developed at birth [11]. The lesions usually increase in size during puberty, and pregnancy, which is due to hormonal changes [12,13]. Alok Bhatt et al. reported a pediatric facial arteriovenous malformation in an 11-year-old female child who presented with pulsatile bleeding, which was treated with embolization and resection [14]. Here in our case, the child presented with asymptomatic swelling over the cheek. Microsurgical resection is the gold standard treatment of all pediatric AVMs [15]. Our child was treated with multiple cycles of sclerotherapy. The most common sclerotherapy agent is Sodium tetradecyl sulphate (STS). It causes endothelial damage by interfering with the cell surface lipids. Finally leads to fibrosis of the lesion with minimal side effects [16]. Some patients complain of discolored urine following sclerotherapy [17]. It is presumed to be due to haemoglobinuria. Rarely it can lead to end-stage renal failure which can be avoided by adequate hydration. The dose of STS depends upon the location and size of the lesion. Our child did not have any side effects. The STS is given as a foam injection. The advantage of foam is, it fills the malformation slowly and causes endothelial damage. Another agent used as the foam is Polidocanol [18]. Our case required multiple cycles of foam sclerotherapy due to the large size of the malformation. The swelling has regressed, and the child is well on follow-up.

4. CONCLUSION

Arteriovenous malformation is not only a physical problem but also involves psychological concern for the child. Though it

may be benign, it might cause facial disfigurement as in our case. A high index of suspicion is required as it might be incidentally diagnosed during dental procedures. Multimodality therapy is necessary for the management and is beneficial in all age groups.

5. ETHICAL STATEMENT

We have obtained written consent from the patient family

6. AUTHORS CONTRIBUTION STATEMENT

Dr Arvinth.S conceptualised and designed the case report. Dr Vindhya. K and Dr Gaddam Aneesh kumar reddy prepared and edited the manuscript. All the authors read and approved the final version of the manuscript.

7. CONFLICT OF INTEREST

Conflict of interest declared none.

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