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Intraosseous venous malformation of the craniofacial region: diagnosis and management

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Abstract

Vascular lesions mainly affect soft tissues, and less than 1% affect bone. In 1982, they were categorised by Mulliken and Glowacki as haemangiomas or vascular malformations, and an updated classification was subsequently published by the International Society for the Study of Vascular Anomalies. These lesions, however, continue to be termed haemangiomas and there is little attempt to differentiate between them. We report eight cases of intraosseous venous malformation that were inappropriately labelled as haemangioma by clinicians, pathologists, and radiologists. We highlight tailored management, and describe the clinical features, results of investigations to aid accurate designation (histological and immunohistochemical, including GLUT1 staining and cross-sectional imaging), and outcomes.

Keywords: Venous malformation; haemangioma; vascular malformation; intra osseous venous malformation; GLUT-1

Introduction

Vascular lesions mainly affect the soft tissues, and less than 1% occur in the bone.[1] In 1982 Mulliken and Glowacki described specific clinical, cellular, and histochemical criteria to classify them into two categories: haemangiomas and vascular malformations.[2] More recently, the International Society for the Study of Vascular Anomalies (ISSVA) has published an updated classification[3] that divides the vascular lesions into vascular tumours or malformations. However, in spite of these well-described multidisciplinary criteria and classifications, vascular anomalies continue to be termed "haemangiomas" and there seems to be little attempt to distinguish them from vascular malformations.[4–6]

Intraosseous vascular anomalies often present in the fourth decade of life and have a female: male ratio of 3:1. They are present at birth and become more obvious later in life, continuing to grow with age.[4] Bone is affected in 35% of cases.[7] Trauma is associated with some, and at least half of those that affect the nasal bone are associated with previous injuries.[8,9]

We document eight cases of intraosseous venous malformation in the craniofacial region, describe their management and outcomes, and highlight their presentation and the investigations done to aid accurate diagnosis.

Methods

We identified all the patients with intraosseous vascular malformations in the craniofacial region who were managed in our unit, and selected those with low-flow venous malformations. Those with high-flow arteriovenous lesions were

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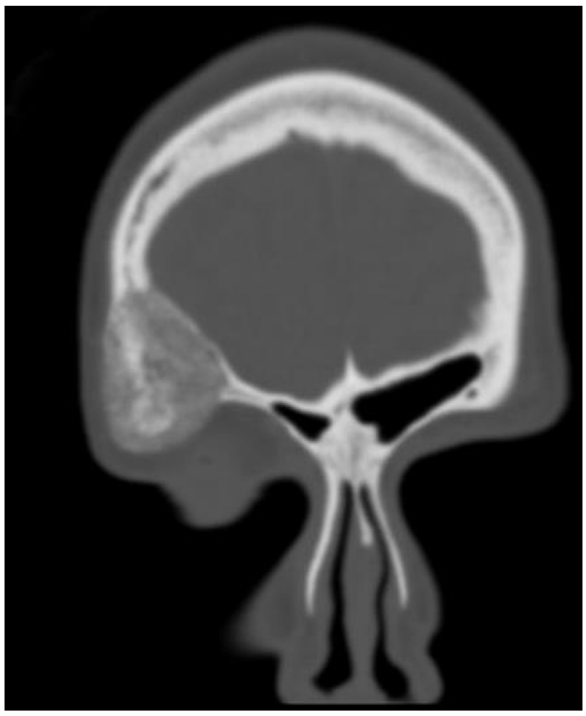

Fig. 1. Coronal computed tomogram showing the lesion arising from the right supraorbital region, with intact cortices and a reticular/radiating pattern of trabeculae.

excluded. The details collected included age, sex, presenting symptoms, history of trauma, clinical features, imaging investigations and characteristics; together with initial clinical, histological and radiological diagnoses, final diagnosis, treatment, and outcomes.

Results

Eight patients with low-flow intraosseous venous malformations were identified (mean age (range) 51 (29–72) years, male:female ratio 1:3). The patients presented with lesions they had had for between one and six years. Four had a history of trauma. Clinical features were mainly of an asymptomatic enlarging mass with discomfort in two. All patients had contrast-enhanced cross-sectional computed tomography (CT) and only one had magnetic resonance imaging (MRI) and a catheter angiogram. CT showed honeycomb/sunburst features in all cases (Fig. 1). Haematoxylin and eosin (H & E) were used for initial stains, and four also had glucose transporter 1 (GLUT1) staining. H & E staining showed widened intertrabecular spaces with dilated vessels that were lined with bland, flattened endothelial cells (Fig. 2). The red cells within the lumen stained with GLUT1, but the endothelial cells in the cell wall did not. The initial histopathological diagnoses were bony haemangioma (n = 6) and cavernous haemangioma (n = 2). None was initially reported as a venous malformation.

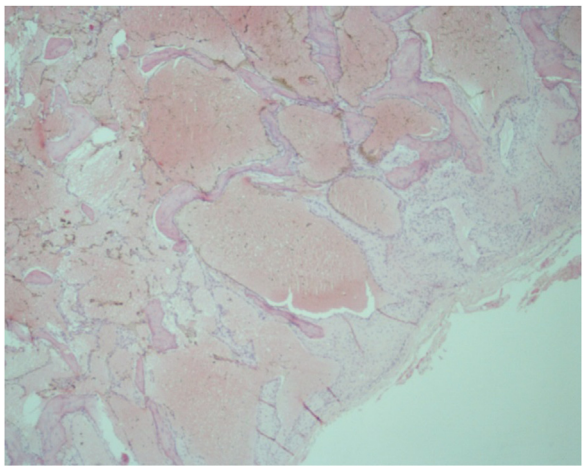

Fig. 2. Photomicrograph showing mature bony trabeculae separated by thin-walled vascular spaces lined by flattened endothelium but no uniform layer of muscle (haematoxylin and eosin, original magnification × 4).

Lesions were excised in seven of the eight patients. Surgical guides were used in two and intraoperative navigation in two. The resulting defects were reconstructed with patient-specific implants in two (polyether ether ketone (PEEK) in one, and custom-made titanium mesh in the other), an iliac bone graft in one, and a composite scapular flap in one. There were no intraoperative or postoperative complications with regards to bleeding or infection. Patients were followed up for between three months and five years, and there was no evidence of recurrence. Detailed results are presented in Table 1.

Discussion

Vascular anomalies mostly affect soft tissues. Primary intraosseous vascular lesions that affect the craniofacial region are uncommon, and account for less than 1% of osseous tumours.[2] Since the seminal work by Mulliken and Glowacki in 1982, vascular lesions have been categorised principally as haemangiomas and vascular malformations.[2,10] They have been further categorised as high-flow and low-flow lesions.[11] The ISSVA published a unified classification system in 1997,[12] and the updated version in 2014[3] divided them into vascular tumours or malformations.

Haemangiomas are characterised by increased cellular proliferation and are largely self-limiting, whereas malformations are characterised by an abnormal development of vasculature, and progress over time.[2–4] Although there has been a widely accepted classification system for vascular lesions for over 30 years, published reports continue to misuse the nomenclature and terminology.[4,13,14] Accurate designation is more than semantic, as the distinct clinical and histological characteristics of the two lesions necessitate appropriately tailored treatment plans and management.[2,3,14,15] Hassanein et al[13] searched for the word

Table 1
Patients' details, investigations, treatment, and outcome.

Case No.	Age (years)/sex	Duration (years)	Trauma	Clinical diagnosis	CT imaging	Site	Initial histological diagnosis	Immunohis-tochemical analysis	Treatment	Reconstruction	Follow up	Recurrence
1	72/F	1	No	Osteoma	Bony haemangioma	Frontal bone	Cavernous bony haemangioma	GLUT- not done	Excision	PEEK implant	1 year	No
2	53/F	3	Yes	Odontogenic cyst	Adenomatoid odontogenic tumour	Anterior maxilla	Haemangioma	GLUT1 negative	Excision	Scapular free flap	5 years	No
3	50/M	6	Yes	Bony haemangioma	Benign lesion/ haemangioma	Parietal bone	Cavernous haemangioma	GLUT1 negative	Excision	None	1 year	No
4	61/F	2.5	INA	Haemangioma	Intraosseous haemangioma	Frontal bone	Haemangioma	GLUT1 negative	Excision	Customised titanium mesh	3 months	No
5	43/F	5	Yes	Myositis ossificans	Bony haemangioma	Zygoma	Haemangioma	GLUT- not done	Excision	Iliac bone graft	10 months	No
6	29/M	4	INA	Haemangioma or sarcoma	Erosion of cortex with sclerotic border	Zygoma	Haemangioma	GLUT- not done	Excision	None	6 months	No
7	66/F	4	No	Exostosis	Benign bony mass	Zygoma	Haemangioma	GLUT negative	Excision	None	2 year	No
8	36/F	4	Yes	Exostosis	Fibrous dysplasia	Frontal bone	Bony haemangioma	GLUT- not done	Incision biopsy	NA	1 year	N/A

"haemangioma" in published reports and found that in 71% (228/320) of cases the terminology had been misused. Of those misdiagnosed, 21% had been treated inappropriately whereas all those with an accurate diagnosis had had appropriate treatment. Precise terminology is essential.

The use of inaccurate nomenclature is even more pronounced in the case of intraosseous vascular or venous malformation.[15–19] In our series all the lesions were initially designated as intraosseous "haemangiomas" and none was described as a venous malformation (Table 1).

Most patients in our series presented with an enlarging hard lump, and two reported associated discomfort. Four of them had a history of trauma, which agreed with findings in other studies.[8,9,20] The age range also agreed with those reported in other studies (29–72 years), though most patients presented after the fourth decade. In the case of arteriovenous malformations, most present in the third decade.[21]

In all cases, contrast-enhanced CT showed a typical sunburst, radiating spoke wheel, reticular, or soapy-bubble appearance, with relatively intact cortices. Though the reported CT features were typical of those described for intraosseous venous malformations,[18] none was designated as such. Instead, four were described as bony haemangiomas, and others as a benign bony mass, or fibrous dysplasia, erosion with sclerotic cortex, or adenomatoid odontogenic tumour.

MRI helps to evaluate the extent of lesions in the soft tissues and it was done in only one of our patients in whom the diagnosis was uncertain. This patient also had a catheter angiogram to evaluate the flow and assess the suitability for preoperative embolisation, but it did not show any appreciable arterial input or vessels that were suitable. Catheter angiograms are seldom required in the diagnosis of an intraosseous venous malformation, and are considered only if they are likely to influence the intervention.[4] None of our patients had the flow evaluated by Doppler ultrasound.

Diagnosis can often be established with a thorough history, clinical examination, and imaging. Three patients had incisional biopsies to establish the diagnosis, and the other five initially had excisional biopsies.

Histopathological analysis (H & E), which showed thin-walled, vascular channels lined by flattened endothelium with scant stroma (Fig. 2) but no uniform layer of muscle, can help to differentiate haemangiomas from vascular malformations. Vascular anomalies are notoriously difficult to diagnose and classify because they have similar histological features and – until recently – there were no specific distinguishing markers.[15,22]

Immunohistochemical analysis, particularly GLUT1staining, can help to confirm the diagnosis. North et al first reported its utility for differentiation between haemangiomas and vascular malformations, and showed that 97% of haemangiomas stained for GLUT1 (sensitivity 95%, specificity 100%), but none of the vascular malformations expressed it (sensitivity and specificity 100%) (Fig. 3). Other authors have subsequently confirmed its utility for differentiation between the two lesions, and principally of those in soft tissue.[10]

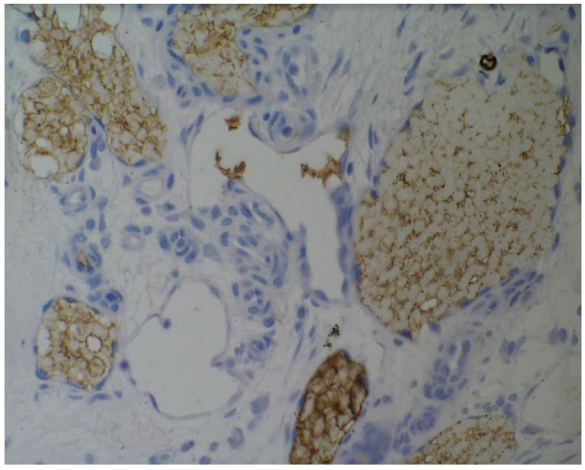

Fig. 3. Photomicrograph showing vessel walls negative for GLUT1 staining (original magnification × 200). The positive cells are the red blood cells, which provide a positive internal control and show that the stain has worked.

In our series, four lesions were stained for GLUT1 but none expressed it. Srinivasan et al[16] and Bruder et al[15] reported the use of GLUT1 in the diagnosis of intraosseous venous or vascular malformations, and highlighted the lack of staining, similar to that seen in soft-tissue lesions. Other authors have subsequently confirmed the lack of GLUT1 staining in intraosseous venous malformations.[17,23]

Operation remains the mainstay of treatment for these lesions. Curettage is generally not recommended because of the risk of bleeding and incomplete excision. Complete excision beyond the perimeter of the lesion is advised to reduce the risk of bleeding and recurrence.[19] Problematic bleeding has not been reported postoperatively when this has been done.[16,19,23] In our series, navigation was used to enable safe and complete excision, and there were no instances of increased intraoperative bleeding.

The resulting defect requires reconstruction to achieve the best functional and aesthetic outcomes. We used patient-specific implants, surgical guides, contoured bone grafts, and scapular free flaps.

Intraosseous vascular lesions continue to be misdiagnosed and a variety of terms is used indiscriminately. Characteristic clinical and imaging features and, where available, histopathological findings including GLUT1 staining, should enable the appropriate categorisation of low-flow lesions such as intraosseous venous malformations. This will enable management to be tailored, and the appropriate evaluation of reports and results.

Ethics statement/confirmation of patients' permission

All patients have given permission for their images and details to be published.

Conflict of interest

We have no conflicts of interest.

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