

Incidental finding of juvenile angiofibroma from pre-orthodontic radiographs: two case reports and a literature review

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ABSTRACT

This case series presents two asymptomatic cases of juvenile angiofibroma which were initially incidentally identified in pre-orthodontic radiographs. Juvenile angiofibroma is an uncommon, locally aggressive benign, vascular neoplasm with invasive growth patterns. Due to the hypervascularity of these tumours, there are biopsy associated risks and multi-slice computed tomography, magnetic resonance imaging and angiography are usually employed for diagnosis. Early pre-symptomatic identification of this lesion facilitates early management and limiting potential life-threatening complications. This highlights the importance of thorough interpretation of dental radiographs, including the evaluation of structures which are not in the primary region of interest. © 2022 Australian Dental Association

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INTRODUCTION

Pre-orthodontic radiographs play an important role in orthodontic assessment, diagnosis and treatment planning. Thorough and accurate interpretation of these images is imperative as incidental findings are considered to be a relatively common occurrence. A study by Tetradis and Kantor reported an average of 1.3 radiographic incidental findings per patient on orthodontic radiographs¹. Moffitt reported that 50% of orthodontists will likely discover significant pathology on lateral cephalograms in their career.²

This case report presents two cases of juvenile angiofibroma (JA) which were initially incidentally identified in pre-orthodontic radiographs. JA is a locally aggressive benign, vascular neoplasm with invasive growth patterns^{3,4}. It commonly occurs in adolescent males who usually present with unilateral nasal obstruction and recurrent epistaxis^{3,5,6}. If asymptomatic, it can remain undiagnosed until late stage. Due to the highly vascular nature of this neoplasm,

biopsies are contraindicated and radiographic diagnosis with computed tomography (CT) and magnetic resonance imaging (MRI) is now the gold standard^{5,7}.

CASE 1

A 15-year-old male was referred by his orthodontist to a private radiology practice for pre-orthodontic radiographs. There were no presenting symptoms and an unremarkable medical history. Anterior displacement of the posterior wall of the left maxillary sinus and widened appearance of the left pterygomaxillary fissure (Fig. 1) were identified by the Oral and Maxillofacial Radiologist and further imaging with CT and MRI was arranged.

These cross-sectional imaging modalities demonstrated a lobulated lesion centred at the widened left pterygomaxillary fissure which is of mixed T2 and low T1 signal on MRI. The lesion extended medially into the posterior nasal space, displacing the nasal septum to the right. It also altered the contour of the nasopharynx and invaded into the sphenoid sinus (Figs 2 and 4). Antero-laterally, the lesion also invaded into the maxillary antrum and masticator

[Correction added on 10 May 2022, after first online publication: CAUL funding statement has been added.]

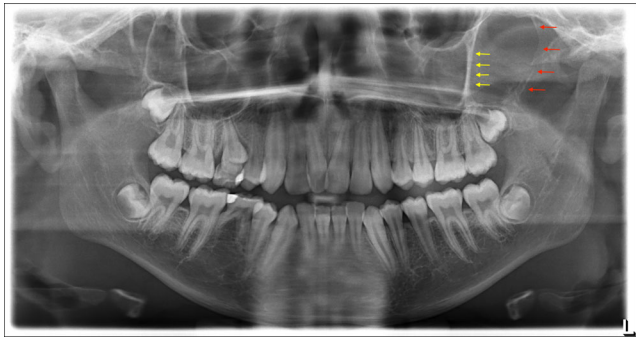


Fig. 1 Case 1 PR image demonstrating anterior bowing of the posterior wall of the left maxillary sinus (yellow arrows) and widened appearance of the left pterygomaxillary fissure (red arrows).

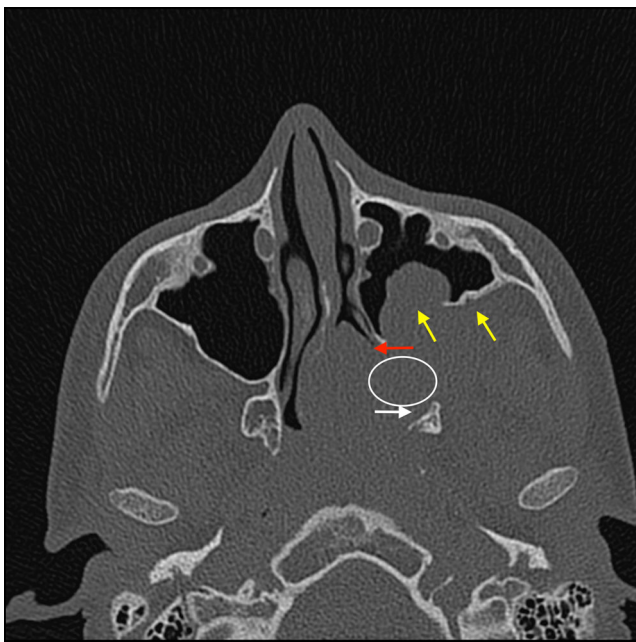


Fig. 2 Case 1 axial CT image demonstrating anterior bowing and erosion of the posterior wall of the left maxillary sinus (yellow arrows), destruction of the lateral wall of the left nasal cavity (red arrow) and destruction of the left pterygoid plates (white arrow). Widening of the sphenopalatine foramen and pterygopalatine fossa (oval area).

space deep to the temporalis muscle, involving the pterygoid muscular complex (Figs 2 and 3). There were no internal calcifications on CT (Fig. 2) and avid contrast enhancement with flow voids in the lesion matrix on MRI (Figs 3 and 4). This was diagnosed radiologically as JA.

The patient was referred to a tertiary hospital and underwent angiographic embolization with surgical removal of the lesion (Fig. 5). The recovery was uneventful.

CASE 2

A 26-year-old male was also referred by his orthodontist to a private radiology practice for pre-orthodontic

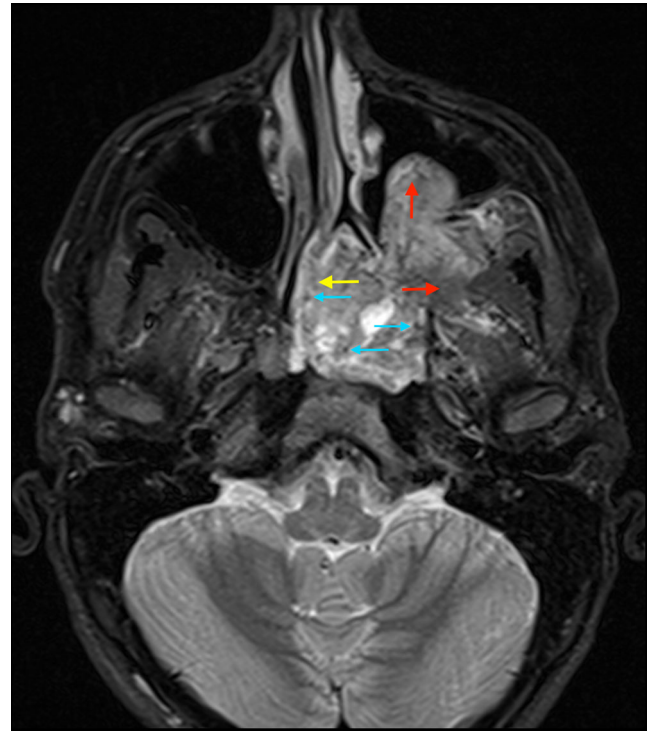


Fig. 3 Case 1 axial T2 weighted MRI image demonstrating anterolateral extension of the tumour into the left maxillary sinus, pterygopalatine fossa, pterygoid musculature (red arrows) and medial extension into the nasopharynx (yellow arrow). Matrix flow voids (blue arrows) and salt and pepper appearance indicative of hypervascularity.



Fig. 4 Case 1 sagittal gadolinium-enhanced T1FS MRI image demonstrating avid enhancement and extension into the sphenoid sinus with no intracranial extension (red arrows).



Fig. 5 Case 1 PR image of the patient post-operatively. Note is made of the embolisation coils and vascular clip present.

lateral cephalogram examination. There were no presenting symptoms and an unremarkable medical history. Anterior bowing of the posterior wall of the maxillary sinus and a large soft tissue prominence at the postero-superior nasopharynx was identified on the lateral cephalogram (Fig. 6) by an Oral and Maxillofacial Radiologist. These appearances were considered strong indicators of a mass lesion, including JA, prompting further imaging.

CT and MRI revealed an avidly contrast enhancing mass in the left posterior nasal space which extensively widened the sphenopalatine foramen and pterygopalatine fossa with scalloping of the maxilla and pterygoid bones extending laterally to the pterygo-maxillary fissure (Figs 7 and 8). On T1 weighted MRI the lesion was isointense with muscle and of mixed signal on T2 weighted imaging, with a number of prominent matrix flow voids (Fig. 8). The lesion insinuated about the ventral lateral pterygoid muscle insertion. Medially there was also ipsilateral septal and posterior turbinate displacement. Superiorly the lesion eroded and bulged into the sphenoid sinus. This was diagnosed radiologically as JA.

The patient was subsequently referred to ENT for further assessment and treatment.

DISCUSSION

The term juvenile angiofibroma was coined by Chauveau in 1906 but the oldest known account is in the 5th Century BC, where written records exist of Hippocrates performing a longitudinal splitting of the nasal ridge to remove a JA lesion⁸⁻¹⁰. The reported incidence is between 0.05% to 0.5% of all head and neck neoplasms with the majority being documented in adolescent males^{4,8,11}. The region of origin of JA is largely accepted as around the posterior nasal cavity but there remains some disagreement with regards to the specific site of origin with primary hypotheses being the sphenopalatine foramen, pterygopalatine fossa, choana and nasopharynx^{8,11,12}.

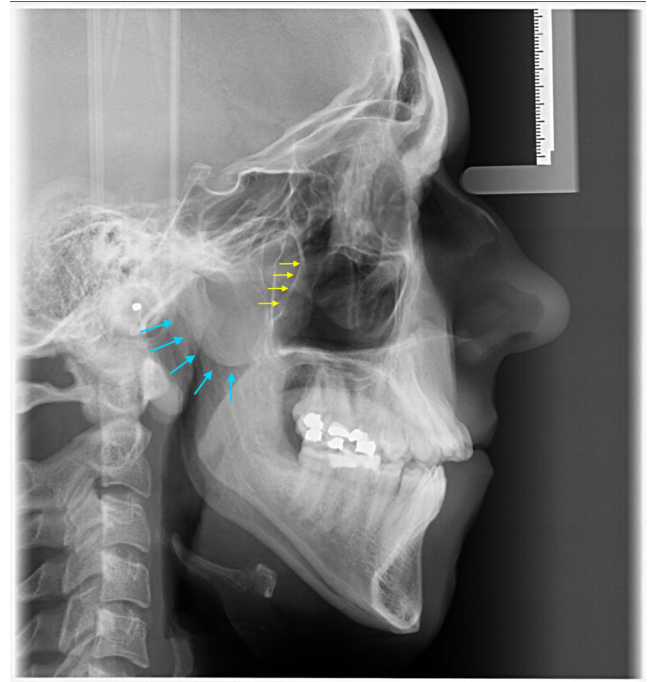


Fig. 6 Case 2 lateral cephalogram demonstrating anterior bowing of the posterior wall of the maxillary sinus (yellow arrows) and the soft tissue prominence within the nasopharynx (blue arrows).

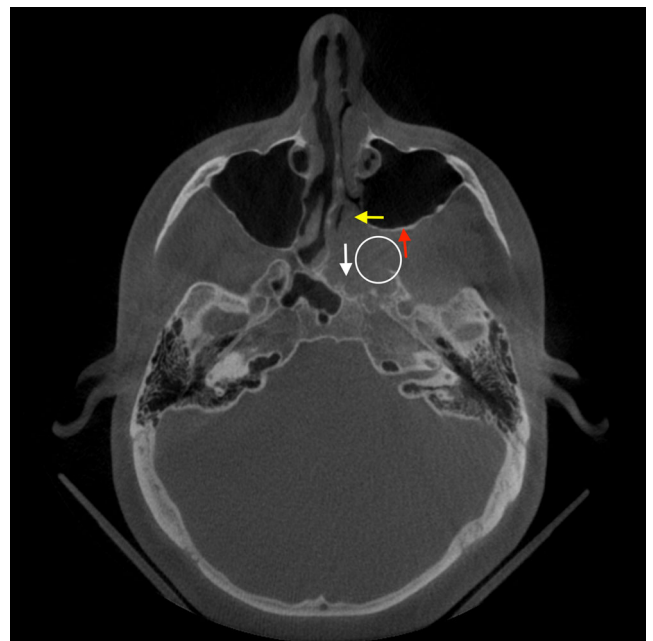


Fig. 7 Case 2 axial CT demonstrating bony changes. Anterior displacement of the posterior wall of the left maxillary sinus (red arrow), destruction of the lateral wall of the left nasal cavity (yellow arrow) and left sphenoid bone (white arrow). Widening of the sphenopalatine foramen and pterygopalatine fossa (oval area).

The aetiology of JA has been disputed and the current perspective is that hormonal, genetic factors and HPV infection could all play a role in its development^{11, 13}. One of the prevailing hypotheses is that JA

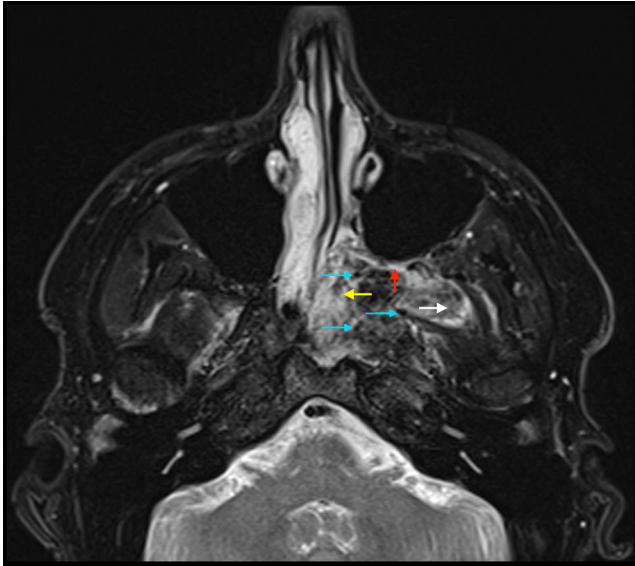


Fig. 8 Case 2 axial T2 weighted MRI image demonstrating soft tissue extensions. Anterior displacement of the left maxillary sinus (red arrow), medial extension into nasopharynx (yellow arrow) and lateral extension into the infra-temporal fossa (white arrow). Matrix flow voids (blue arrows) and salt and pepper appearance indicative of hypervascularity.

is a vascular malformation derived from remnants of the first branchial arch artery. The angiofibromatous tissue is likely triggered to proliferate due to excess sex hormone receptors being present on normal nasal mucosa⁵. Multiple sex hormone receptors have been isolated from JA lesions including androgen receptors, oestrogen receptors α , follicle-stimulating hormone receptors and luteinizing hormone receptors⁷. These receptors and their surplus are triggered during puberty due to peak fluctuations in sex hormones⁵, which could explain the male predilection. Case 1 demonstrates the more typical identification in an adolescent male. Case 2 involved a young adult male although it is suspected that the lesion had likely been present for some time.

JA is a benign neoplasm but behaves aggressively showing local expansion along planes of least resistance. It is associated with bony erosion and destruction of cortical boundaries and has a propensity to spread through foramina and fissures^{5,6}. Most commonly patients present with complaints of unilateral nasal obstruction and recurrent epistaxis¹⁴. Other symptoms such as proptosis, hypernasal speech and cranial nerve deficits are indicative of larger lesions⁵. The case series presented here demonstrates that cases could present as an incidental radiographic finding.

Diagnostic protocol for JA has changed in recent years with the advancements in imaging. Biopsy has fallen out of favour and is now contraindicated due to the risk of uncontrollable haemorrhage^{5,7,10,13,15-17}. Diagnosis and staging of lesions are now based on advanced imaging modalities using CT, MRI and angiography. Lesions can be accurately mapped where

CT is used to demonstrate bony changes, for surgical planning and now can be applied to image-guided surgery⁷. The imaging features on CT are a non-encapsulated mass with heterogeneous density which is commonly located around the sphenopalatine foramen and pterygopalatine fossa. It tends to expand medially through the nasopharynx and nasal cavity, anterolaterally to displace the posterior wall of the maxillary sinus and cause widening of the pterygopalatine fossa⁶. This was demonstrated in both cases, where widening of the pterygopalatine fossa was evident, with more prominent anterolateral extension in case one. Superiorly, it can extend into the skull base through foramina and fissures but intracranial extension is only noted in 10-20% of cases¹³. Superior extension was evident in both cases with bulging into the sphenoid sinus but no intracranial extension.

MRI is superior in assessing soft tissue and intracranial extension and for demonstrating intra-lesional signal voids which are a feature of JA lesions due to their hypervascularity^{3,5,17}. MRI is also important in delineating the lesion from important surrounding structures such as blood vessels, nerves, dura and marrow⁷. The imaging features on MRI are an isointense to hyperintense lesion on T1 and T2 weighted scans with multiple intra-lesional signal voids⁷. Lesions enhance intensely with IV gadolinium as seen in this case series⁷. Angiography is used to assess the lesion's vascularity, including its arterial blood supply, venous drainage and internal vascular composition¹⁸. The characteristic imaging features of JA allows accurate diagnosis, eliminating the need for a treacherous biopsy of a vascular lesion. The imaging modalities discussed result in accurate mapping of lesions that are vital in their management and surgical intervention.

Management of JA involves surgical resection with pre-operative angiographic embolisation and post-operative radiotherapy as adjuncts to consider. Pre-operative embolisation of feeder vessels reduces the risk of bleeding complications during surgery^{13, 17, 19}. Case one in this series was managed successfully with angiographic embolisation and surgical resection. Radiotherapy application is used in cases of residual, recurrent disease or if the surgical morbidity risk is high^{13,17,19}. Image-guided surgery with the use of CT imaging is being increasingly employed to improve the accuracy of resection and surgical outcomes⁷. Management modalities applied to a case are dependent on the size, extension, location and staging of the lesion¹⁹.

CONCLUSION

Juvenile angiofibroma is a benign but locally aggressive neoplasm of the maxillofacial region. Although patients usually present with symptoms, the two cases described represent asymptomatic lesions which were

incidentally identified in pre-orthodontic radiographs, facilitating early diagnosis and treatment. This highlights the importance of accurate interpretation of imaging commonly performed in dentistry, including the thorough evaluation of structures beyond the regions of interest. Due to the vascular nature of juvenile angiofibroma, radiologic assessment is now the gold standard and surpasses biopsy and histopathology for diagnosis. Advancements in CT and MRI imaging has also allowed highly accurate mapping of lesions which is crucial in their surgical management and long term follow up.

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