International Journal of Paediatric Dentistry 2006; **16:** 302–306

Rhabdomyosarcoma of the mandible in a 6-year-old boy

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Summary. *Introduction.* Rhabdomyosarcoma is an aggressive malignant tumour composed of neoplastic mesenchymal cells that infiltrate surrounding tissue structures, making their precise site of origin unclear. Although rare, this is highly aggressive and the most common soft-tissue neoplasm of the head and neck in children. Regrettably by the time most cases are initially seen, the patients already have large tumours, due to rapid tumour growth and delayed medical consultation.

Case Report. This report describes a 6-year-old presenting with just such symptoms of facial swelling and pain but elicitation of further information and findings, including tooth mobility of 3 days duration, led to prompt referral and early treatment of an embryonal rhabdomyosarcoma.

Conclusion. General dental practitioners are frequently presented with a child with a swollen face and pain. Experience would suggest a dental abscess to be the most likely cause with treatment as appropriate. However, all swellings in children, should be thoroughly investigated and reviewed as particularly in this age group, tumour growth is rapid while early diagnosis allows successful treatment with multimodality therapy.

Introduction

It is the duty of each general dental practitioner (GDP) to recognize and manage oral disease. The treatment of any condition can only be effective if the aetiology of the problem has been established. Initial diagnosis is usually made after history and examination, with special tests if appropriate based on a knowledge of disease processes and experience.

The possible causes of facial swellings in a 6-year-old child include odontogenic infection, most commonly dental abscess, cellulitis, or pericoronitis, as well as the less common nondental infections such as sialadenitis, atypical mycobacteria, and osteomyelitis [1]. Neoplasms, usually benign, may occur, while malignancies should always be considered. Alerting features to the latter should be painless swelling, tooth mobility, and/or extrusion and unusual radiographic appearance.

Rhabdomyosarcoma is the most common malignant soft-tissue tumour in children representing approximately 4–8% of all cases of malignant disease in those younger than 15 [2]. Although 40% of rhabdomyosarcomas are located in the head and neck region, there would appear to have been no previous reports with the mandible as the site of origin. Early diagnosis is crucial as the disease, previously fatal, can now be successfully treated with multimodal therapy [3].

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Case report

A 6-year-old Caucasian boy had visited his dentist the previous month for a check-up at which time no abnormality had been noted. Subsequently 4 weeks later he attended the same practitioner as an emergency patient complaining of a right-sided facial swelling of 3 days duration. The history elicited was of an apparently symptomless intraoral swelling of 2 weeks duration for which no professional advice had been sought. The practitioner noted the first lower right permanent molar to be extruded and had grade III mobility. An orthopantomo graph was undertaken and revealed a ‘diffuse’ area at the angle of the mandible. This combination of clinical and radiographic findings led the GDP to refer the patient urgently by telephone to the Paediatric Dentistry Clinic when he was seen later the same day.

The child’s obstetric history was delivery via an emergency caesarean section at term and a week. Developmental milestones and immunizations were all on target. With the exception of an overnight admission in 1999 with ‘probable pneumonia’ the medical history was uneventful.

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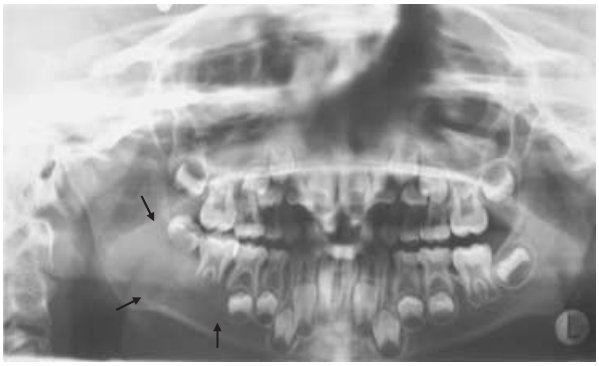

Fig. 1. OPT showing extrusion of 46 and 47 with loss of lamina dura and ID canal destruction.

On examination there was a marked facial swelling in the right parotid region together with submandibular and cervical lymphadenopathy. The swelling was neither warm nor tender and the overlying skin was of normal colour. Paraesthesia was noted in the right lower lip and the patient reported his 'poor lip' as having felt different for 2 weeks.

Intraorally, the mandibular right first permanent molar was extruded 4 mm and lingually inclined with grade III mobility. The gingivae were heaped over the disto-buccal surface.

The OPT radiograph provided by the GDP (Fig. 1) showed the mandibular right first permanent molar to be extruded with loss of the lamina dura. The mandibular right second molar appeared vertically displaced. There was loss of clarity of the entire crypt of mandibular right first and second molars and the mandibular right second premolar distally. The right inferior dental canal was indefinable.

A computed tomography (CT) scan of the mandible together with the head and chest was arranged for the following day at Sheffield Children's Hospital, because the child was highly cooperative this was undertaken without sedation or general anaesthesia (GA). The scan revealed a large ill-defined lytic lesion within the ramus and angle of the right mandible (Fig. 2). Destruction of the lamina dura of the mandibular right first and second molar was present. Spicules of calcification extended into the soft tissue adjacent to the lesion. The soft-tissue mass associated with the bony destruction could not be separated from the masseter or the medial pterygoid muscles on the right side. The neck, mediastinum, lungs, and upper abdomen appeared normal. A bone scan showed abnormality in the site of the lesion only.

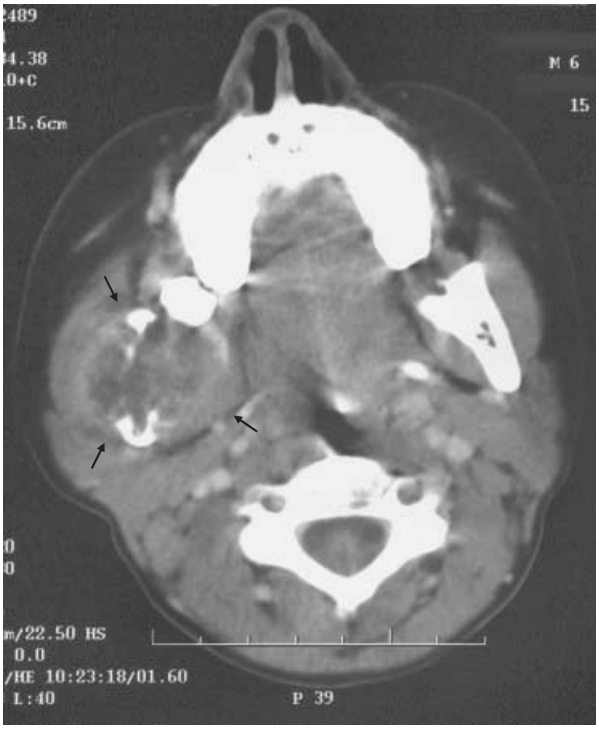

Fig. 2. CT scan showing large, destructive lytic lesion within the ramus and angle of the right mandible. Spicules of calcification extend into the associated soft tissue mass.

On the same day, the patient was admitted to Sheffield Children's Hospital for treatment under GA. This included an incisional biopsy in the lower right molar region together with removal of unerupted lower right second permanent molar by the oral and maxillofacial surgery team. The biopsy specimens were sent for histological and immunohistochemical analysis. Blood samples taken for full blood count, urea and electrolytes, liver function tests, uric acid, and serum lactose dehydrogenase (LDH) were all within normal limits as was a hearing test subsequently undertaken. The histology reported the appearance of a small round cell neoplasm with variable mitotic activity consistent with a sarcoma.

Management of the tumour was planned via a multidisciplinary approach with induction chemotherapy to shrink the margins prior to definitive surgical resection followed by further consolidation chemotherapy. It was thus hoped to avoid the complications of radiotherapy if complete surgical resection could be obtained.

Placement of a Broviac line under general anaesthesia was undertaken to facilitate the administration

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of the six courses of chemotherapy agents (vincristine, ifosfamide, doxorubicin, etoposide – VIDE) each requiring a 2-night hospital admission at 3-week intervals. During this period, the patient re-presented with febrile neutropenia on three occasions and tongue pain, mouth ulcers, upper respiratory tract infection and mandibular swelling once each on separate occasions.

Following induction chemotherapy, there was a detectable reduction in tumour size visible from specialist scanning. There was no regional lymphadenopathy and surgery was undertaken 5 months after initial presentation. This included a tracheostomy, lip split, segmental resection of the mandible extending from lower right permanent second premolar to include condyle and coronoid processes with resection of the adjacent masticatory muscles and neck dissection at levels 1 and 2. Reconstruction with a fibular osseoseptalcutaneous free flap was performed with the assistance of the plastics team.

Unfortunately, the initial surgery was complicated by failure of the graft within the first 24 h because of vascular problems. This was subsequently removed and replaced a few days later with a costochondral rib graft. The child made an excellent recovery with a 3-day stay in intensive care and initial period of nasogastric feeding. The tracheostomy was removed 9 days following placement. This entire admission was just over 2 weeks and the only minor problem related to the infection of his leg wound that was treated with antibiotics. Over a 2-month period, rehabilitation with the use of crutches and physiotherapy helped to resolve the initial limp.

Histological examination of the resected specimens showed the margins to be clear of tumour. The tumour appeared to have arisen within the mandible as macroscopic examination demonstrated total replacement of the cancellous bone and marrow spaces with destruction of the cortical bone, but only limited extension into soft tissue. The immunostaining patterns were suggestive of an embryonal rhabdomyosarcoma but because of the origin in bone the descriptive diagnosis of sarcoma with a rhabdoid differentiation resembling an embryonal rhabdomyosarcoma was deemed more appropriate.

Less than 2 weeks after discharge following surgery the first of eight further courses of chemotherapy (vincristine, actinomycin, ifosfamide – VAI) were commenced requiring the same admission pattern.

Gabapentin was prescribed 2 months after surgery for vincristine-related jaw pain and proved effective. Admission was necessary around the fifth course of post-surgery chemotherapy because of history of fever and inflammation around the scar. This resolved with intravenous antibiotics. Following the seventh course of chemotherapy, admission was required because of a line infection that resolved with intravenous antibiotics. Following satisfactory follow-up scans, the Broviac line was removed.

The patient's treatment was complete 11 months after initial presentation and he has subsequently undergone booster vaccinations as per the Royal College of Paediatrics and Child Health guidelines [4] 6 months after completion of treatment. Overall, he coped extremely well with all aspects of his treatment both physically and psychologically with no other oral problems encountered.

The patient has been reviewed on average of twice monthly over the last 14 months since completion of treatment and remains well within himself and has excellent jaw movement (Figs 3, 4, 5). He receives shared care with his GDP as part of his family unit and at the dental hospital where his dentofacial development is monitored at appointments coincident with medical follow-up. Preventive advice and

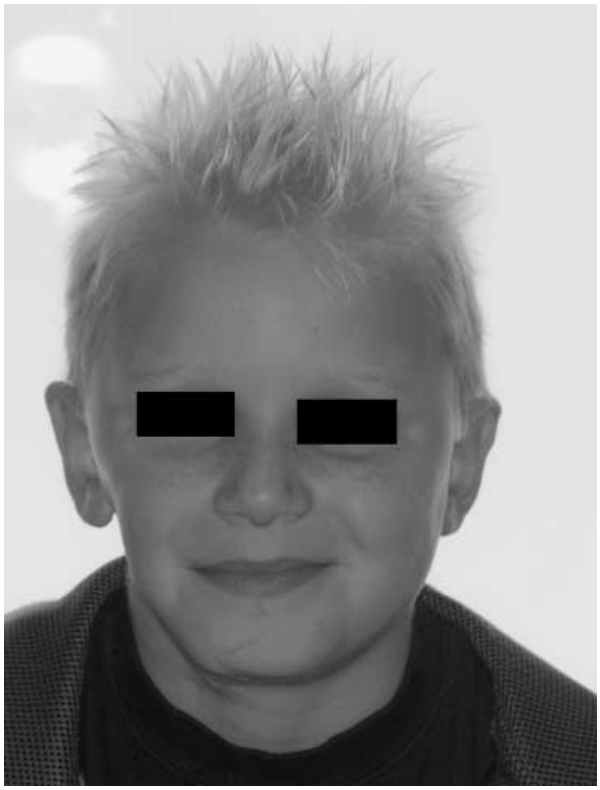

Fig. 3. Facial view following reconstructive surgery.

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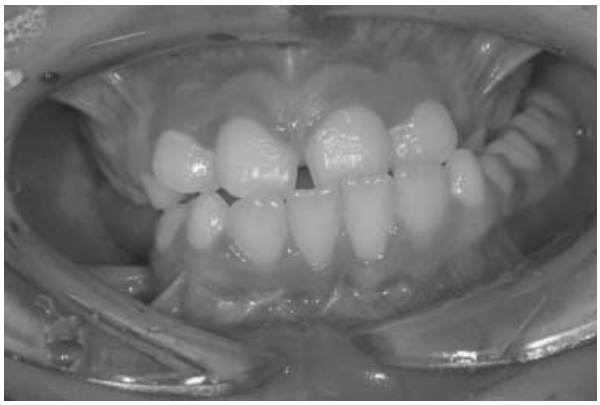

Fig. 4. Intraoral view showing developing occlusion.

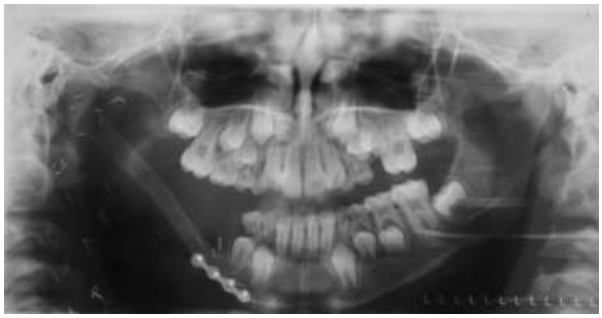

Fig. 5. Radiograph illustrating costochondral rib graft *in situ*.

treatment including dietary advice, oral hygiene instruction, placement of fissure sealants, topical fluoride application, and prescription of chlorhexidine rinse when necessary are provided at both centres.

Discussion

Rhabdomyosarcoma (RMS) is an aggressive malignant tumour composed of neoplastic mesenchymal cells, with varying degrees of striated muscle differentiation. It has rapidly growing local extensions including bony destruction. Described as being the most common soft tissue sarcoma in children, it represents approximately 4–8% of all cases of malignant disease in children younger than 15 [2] and 40% occur in the head and neck region [3].

RMS of the head and neck often presents with vague symptoms that mimic other disease conditions. These may include painless swelling, throat discomfort, or nasal airway obstruction [5], factors that may lead to undue delay in the establishment of the correct diagnosis. Other symptoms such as progressive trismus, pain and paraesthesia [6] are more likely to produce a more rapid response. Delay in establishment of the correct diagnosis, however, is reported as ranging from 1 to 17 months with consequential delay in treatment and less successful outcome [7].

Prognosis is directly related to the stage of the disease at diagnosis, anatomical localization, histological type, and degree of tumour anaplasia [8,9]

Since the introduction of the Intergroup Rhabdomyosarcoma Studies (IRS) protocols for the management of RMS, the studies have shown that multimodality treatment involving chemotherapy, surgery, and radiotherapy can now achieve 2-year survival rates of up to 80% at all sites [10]. Complete surgical resection offers the best prognosis and allows the avoidance of radiotherapy [11] with its associated side effects. The effects primarily attributed to radiotherapy for the treatment of RMS of the head and neck in children were recently reported [12]. These include neuroendocrine, thyroid, dental, and cognitive sequelae. In the review of 30 cases a few, but severe, late effects were found including chondronecrosis, oesophageal stenosis, secondary malignancy, and brain haemorrhage.

Not all sites, however, are amenable to surgery alone. Chemotherapy is commonly used to prevent metastases and for elimination of gross or microscopic disease. Previous studies show the most common sites of RMS in the head and neck to be the orbit and nasopharynx [13]. Long-term follow-up of 469 children with localized, non-orbital soft-tissue sarcomas showed a 5-year survival of 53%, thus faring better than adults. It has been proposed that the adjuvant therapy may be contributory [14].

In the case described, the GDP was the first professional to assess the patient and prompt referral allowed complete resection of the localised disease providing optimum prognosis for this young boy. The role of the paediatric dental team is not only to place the patient with the appropriate specialties but also implement preventive regimes, including the placement of fissure sealants, restorations as appropriate, and extractions of teeth of poor prognosis. This in turn will avoid the need to deal with dental emergencies particularly while the patient is undergoing or recovering from chemotherapy or surgery as intensive oncology treatment can induce oral mucositis, neutropenia, and thrombocytopenia, which can make dental treatment difficult.

Estilo *et al*. reported some 80% of RMS patients were found to have clinical or radiographic dentofacial

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abnormalities that can affect their quality of life and proposed early input from dental teams [15]. Disturbance of dental development is well reported [14]. The consequences can include enamel defects, microdontia and incomplete tooth formation with foreshortened and/or blunted roots and even tooth agenesis [16]. This, together with compromise of jaw growth and trismus [15,17], may in turn impact on the feasibility of orthodontic therapy later in life. Again, the practitioner may need to refer for specialist multidisciplinary advice regarding the dental defects, tooth position and facial development.

The importance of the vigilant dental professional must not be underestimated and the need to provide optimal care for both the immediate and long-term for these patients is emphasized especially in view of the improved survival rates.

What this paper adds

- To the very best of our knowledge this paper presents the first report of rhabdomyosarcoma originating in the mandible of a 6-year-old child.
- The possible causes of facial swelling in children are discussed.

Why this paper is important to paediatric dentists

- The management of this unusual case is presented in relation to clinical presentation, investigations and treatment thus reflecting the patient journey.
- The role of paediatric dentistry interface with other specialties, as well the immediate and long-term orofacial needs, is discussed.

Acknowledgements

The assistance from both Dr Paula M. Farthing (Consultant Oral Pathology) and Mr Jeremy McMahon (Consultant Oral and Maxillofacial Surgery) is greatly appreciated.

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