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Pathology Spot

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Rhabdomyosarcoma of the Adult Nasopharynx

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Key Words
Rhabdomyosarcoma · Nasopharynx · Adult

Abstract
Rhabdomyosarcoma of the adult head and neck is rare, particularly beyond 40 years of age. In this region, the nasopharynx is an unusual site. Most nasopharyngeal rhabdomyosarcomas are of an alveolar variety. We report a case of embryonal or spindle cell rhabdomyosarcoma in the nasopharynx of a 47-year-old man. The histology of this tumour revealed clear cells that have not been described in embryonal rhabdomyosarcomas occurring in the head and neck. The pathology is discussed and current literature reviewed.

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Introduction

Rhabdomyosarcoma is a rare neoplasm of the adult head and neck. The tumour produces symptoms such as nasal congestion or otorrhoea, which mimics benign disease states [1]. It often presents and is diagnosed late. Histological misinterpretation of microscopic findings can lead to further delay, as 'cross-striations' which are the hallmark of rhabdoid neoplasms are not always found. Survival for this type of tumour is poor, with an overall survival rate (Kaplan-Meier method) of 7.6% at 5 years [2]. All long-term survivors belong to early-stage groups who respond to a combination of surgery, radiation and chemotherapy. This highlights the importance of an early definitive histological diagnosis.

In the adult head and neck, rhabdomyosarcomas tend to occur before the age of 40 [3]. The paranasal sinuses are the most common primary sites with tumours of the nasopharynx being rare. Histologically, adult rhabdomyosarcomas tend to be of a pleomorphic variety. However, pleomorphic rhabdomyosarcomas are exceedingly rare in the head and neck region occurring more commonly in the extremities. In the nasopharynx, alveolar rhabdomyosarcomas tend to predominate.

We present a case of an embryonal rhabdomyosarcoma in the nasopharynx of a 47-year-old man. The tumour had an abundance of clear cells which have yet to be described in embryonal head and neck rhabdomyosarcomas. This proved to be a histopathological diagnostic challenge.

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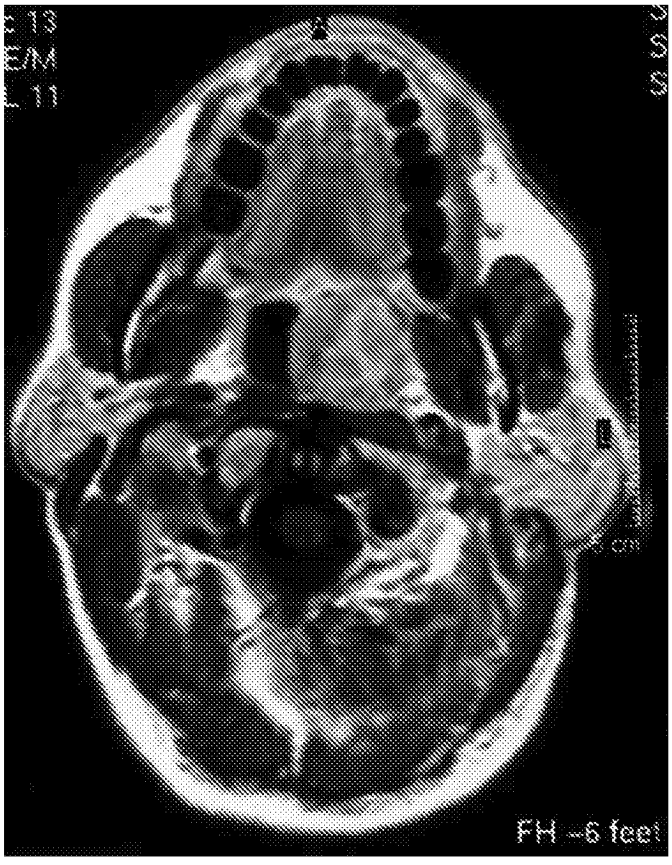

Fig. 1. Axial section T1-weighted MRI scan revealing a large mass occupying the post-nasal space.

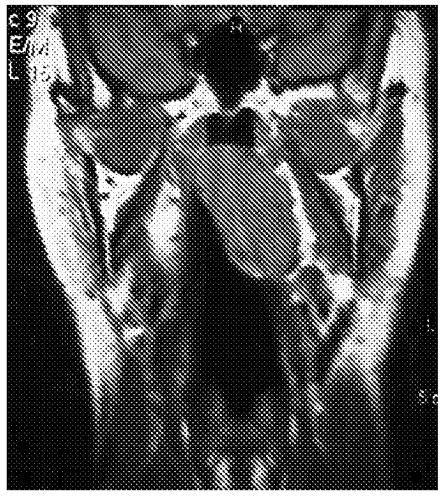

Fig. 2. Coronal section T1-weighted MRI scan showing the well-circumscribed tumour arising from the left lateral wall of the nasopharynx.

Case Report

A 47-year-old Caucasian-male presented with a 12-month history of worsening nasal congestion. He had received treatment for allergic rhinitis with little effect. In the 9 months prior to presentation, he developed symptoms of obstructive sleep apnoea, hyponasality and left-sided hearing loss. He was a life-long non-smoker and consumed 28 units of alcohol a week.

As a child, he underwent an uncomplicated adenoidectomy and tonsillectomy. He had no relevant past medical history other than hypercholesterolaemia for which he took a statin. He gave a family history of colorectal cancer in his father and one sibling, both occurring after the 5th decade.

Examination by nasendoscopy revealed a large bosselated post-nasal space mass displacing the soft palate forward. No neck nodes were present and physical examination was otherwise normal. CT scans showed a well-circumscribed mass arising from the left lateral wall of the nasopharynx (fig. 1, 2). He subsequently underwent resection of the tumour both endoscopically through the nose and from the oropharynx with retraction of the soft palate. The tumour appeared well demarcated and was removed macroscopically in toto. Four quadrants of further tissue excised from the exposed nasopharynx revealed one to be positive for tumour. Further staging investigations were all unremarkable including CT scans of the thorax and abdomen, bone scan, bone marrow aspirate and liver ultrasound.

The patient was then commenced on chemoradiation involving 9-week cycles of actinomycin D and vincristine for 6 months. The initial radiotherapy field included the skull base, post-nasal space and neck.

Pathological Findings

Microscopy of the tumour reveals a mixture of cells. There are sections clearly demonstrating rhabdoid differentiation with elongated strap-like cells lacking cross-striations (fig. 3). These spindle cells have markedly pleomorphic hyperchromatic nuclei. In some cells, the nucleus is located at one end giving the characteristic 'tadpole' or 'comet' shaped appearance. There are also areas showing rounded cells and larger rhabdomyoblast-like cells in nodules with clear cytoplasm (fig. 4). There are occasional but not plentiful mitoses and a focally marked inflammatory infiltrate.

Immunostaining was strongly positive for desmin and vimentin but negative for cytokeratin, GFAP, SMA and MyoD1. There was, notably, some focal positivity for myoglobin.

The abnormal elongated strap like cells clearly suggest striated muscle differentiation. The results of immunostaining confirm a rhabdomyosarcoma. The tumour is too pleomorphic for a rhabdomyoma. The substantial spindle-cell component and lack of alveolar architecture make

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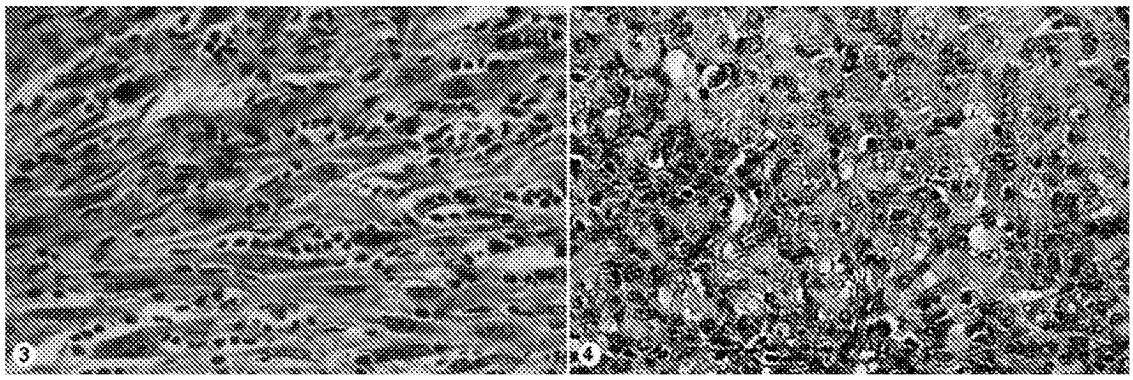

Fig. 3. Elongated strap-like cells suggesting rhabdoid differentiation. There is a lack of cross-striations. HE. × 100.
Fig. 4. Foam cells with abundant clear cytoplasm, but no 'alveolar' structure. Clear cells have not been described in embryonal rhabdomyosarcomas of the nasopharynx. HE. × 100.

further sub-division as alveolar rhabdomyosarcoma inappropriate. Taken as a whole, the histological features of this tumour best fit a diagnosis of embryonal or spindle cell rhabdomyosarcoma of the nasal cavity. The presence of clear cells is an outstanding feature.

Discussion

Rhabdomyosarcoma was first described by Weber in 1854 and is a tumour derived from mesenchyme. It is the third most common sarcoma but is the most common in children in whom they account for half of all cases [4]. In children, the head and neck is the most common site. By contrast, head and neck rhabdomyosarcoma is rare in adults and exhibits a different natural history, response to treatment and prognosis [5].

Macroscopically, these tumours appear soft, fleshy and pink with no distinction between types except for the botryoid variant. This variant appears like a bunch of grapes (from the Greek 'Botrys', a cluster of grapes). Microscopically, the tumour cells are the 'neoplastic analogue of skeletal muscle embryogenesis' [6]. The rhabdomyoblast recapitulates normal development from a primitive round cell to spindle cell to multinucleated muscle fibres, but in a highly disorganised manner. There are four histologic varieties – embryonal, alveolar, pleomorphic and undifferentiated.

Some authorities have proposed that there are only two true varieties; the juvenile form, encompassing the embryonal and alveolar variants, and the adult pleomorphic form.

The cells in embryonal rhabdomyosarcoma resemble those of a normally developing skeletal muscle of a 7- to 10-week fetus [7]. The predominant cell is spindle-shaped and the nucleus, usually single, is located centrally. The cytoplasm is abundant and eosinophilic. Alveolar rhabdomyosarcoma resembles the hollow-tube stage of fetal muscle development normally seen at 10–12 weeks. Small rounded cells grow in bands around central clear spaces resembling 'alveoli'. Pleomorphic rhabdomyosarcoma represents dedifferentiation of normal muscle, producing a spindle cell tumour. They are the commonest variety in adults, but uncommon in the head and neck.

Clear cells, which have a foamy cytoplasm, appear abundantly in our case. They have been described in alveolar rhabdomyosarcoma of the nasal cavity. However, the spindle-cell component, as seen in figure 3, and the lack of a definitive alveolar structure makes this diagnosis unlikely. Whilst embryonal rhabdomyosarcoma provides the best-fit diagnosis, clear cells have never been described in this entity. Mixed alveolar and embryonal rhabdomyosarcoma are occasionally seen in children but are exceedingly rare in adults.

Recent studies [3, 8] have revealed no correlation between histology of tumours and prognosis, contrary to the belief that alveolar tumours fared worse than embryonal ones [9]. In a series of adult head and neck rhabdomyosarcoma [2], nodal metastases were seen in half of all patients at presentation. These patients were in grade III or IV, supporting the hypothesis that nodal involvement is related to primary tumour size [10]. Systemic metastases were present in a fifth of patients. The lung is said to be the most common site [3] although there was a

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noticeable predilection for bone in one series [2], underlining the importance of bone scans in the initial work-up.

In the adult head and neck, complete surgical excision has been shown to be a favourable prognostic factor [11]. Site is therefore an important determinant of survival in so far as it determines resectability. Parameningeal tumours, which cause bone erosion, are associated with poorer prognosis [12, 13].

Chemoradiation following complete surgical resection appears to be the therapeutic ideal. This is based on success rates in children who have a 71 and 59% 4-year survival rate for non-parameningeal and parameningeal tumours, respectively [14]. As the biologic properties of adult head and neck rhabdomyosarcoma are different and the tumour rare, there is little evidence to support any particular therapeutic programme.

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