

Embryonal Rhabdomyosarcoma of the Nasal Cavity in an Adult : A Case Report

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Embryonal rhabdomyosarcoma of the head and neck is very rare in adults. We report a case of embryonal rhabdomyosarcoma in the nasal cavity, occurring in a 24-year-old male patient presenting with left nasal obstruction, ipsilateral eye and cheek pain and multiple neck nodules. Rhabdomyosarcoma should be considered in the differential diagnosis of poorly defined soft tissue mass of the nasal cavity not only in children, but also in adults.

Index words : Embryonal rhabdomyosarcoma
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Rhabdomyosarcoma is the most common soft-tissue sarcoma that is most typically found in children under 15 years of age (1). The World Health Organization distinguishes predominantly pleomorphic, alveolar, embryonal and mixed types of rhabdomyosarcoma (2). Head and neck rhabdomyosarcoma is common in childhood, however, is very rare in adults, especially of the embryonal type in the nasal cavity (2, 3). Soft tissue sarcoma makes up less than 1% of all adult malignancies, and rhabdomyosarcoma accounts for 3% of all soft tissue sarcomas (4). Sultan et al. (5) reported that only 9.3% of all adult rhabdomyosarcomas occurred in the head and neck region.

A case of rhabdomyosarcoma of the embryonal type located in the nasal cavity of a 24-year-old male is presented and the literature on CT and MR appearance is reviewed.

Case

A 24-year-old man presented with left nasal obstruction, ipsilateral eye and cheek pain and multiple neck nodules for two months. Clinical examination revealed a mass in the left nasal cavity. Computed tomography (CT) and magnetic resonance imaging (MRI) of the nasal cavity and paranasal sinuses were performed.

CT scan with a bone window setting showed a poorly defined, relatively isodense with adjacent muscle, heterogeneous mass of the left nasal cavity that destroyed adjacent bony structures and extended to the surrounding spaces, such as ipsilateral ethmoidal sinus, orbit, nasopharynx, maxillary sinus, sphenoidal sinus, foramen lacerum, cavernous sinus and sphenopalatine foramen. After intravenous administration of contrast material, the mass enhanced to the same degree as adjacent muscles (Fig. 1). MR imaging showed isointense signal in-

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tensity of the mass with adjacent muscles on T1-weighted images and slightly high signal intensity on T2-weighted images. After administration of contrast material, the mass enhanced more than adjacent muscles. Trapped secretion between the mass and the sinus wall was distinguishable from tumor; it was slightly hy-

pointense on the T1-weighted sequence and hyperintense on the T2-weighted sequence and did not show contrast enhancement. Nodal extension was demonstrated in the both carotid, retropharyngeal and left submandibular spaces (Fig. 2). Calcification or intratumoral hemorrhage was not found in this case.

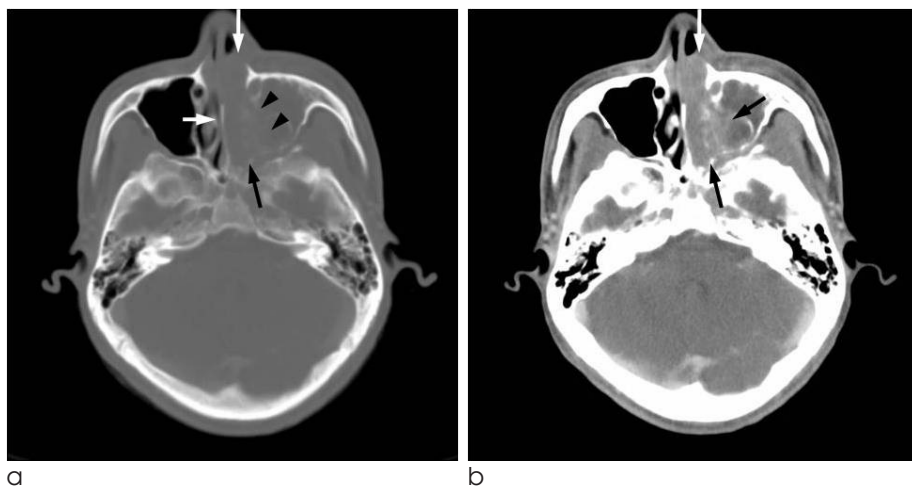


Fig. 1. a. Axial CT scan with bone window setting shows a poorly defined, heterogeneous, relatively isodense mass of the left nasal cavity. The mass destroys adjacent bony structures and extends to the ipsilateral maxillary sinus. b. On contrast-enhanced axial CT scan, the mass is enhanced to the same degree as adjacent muscles.

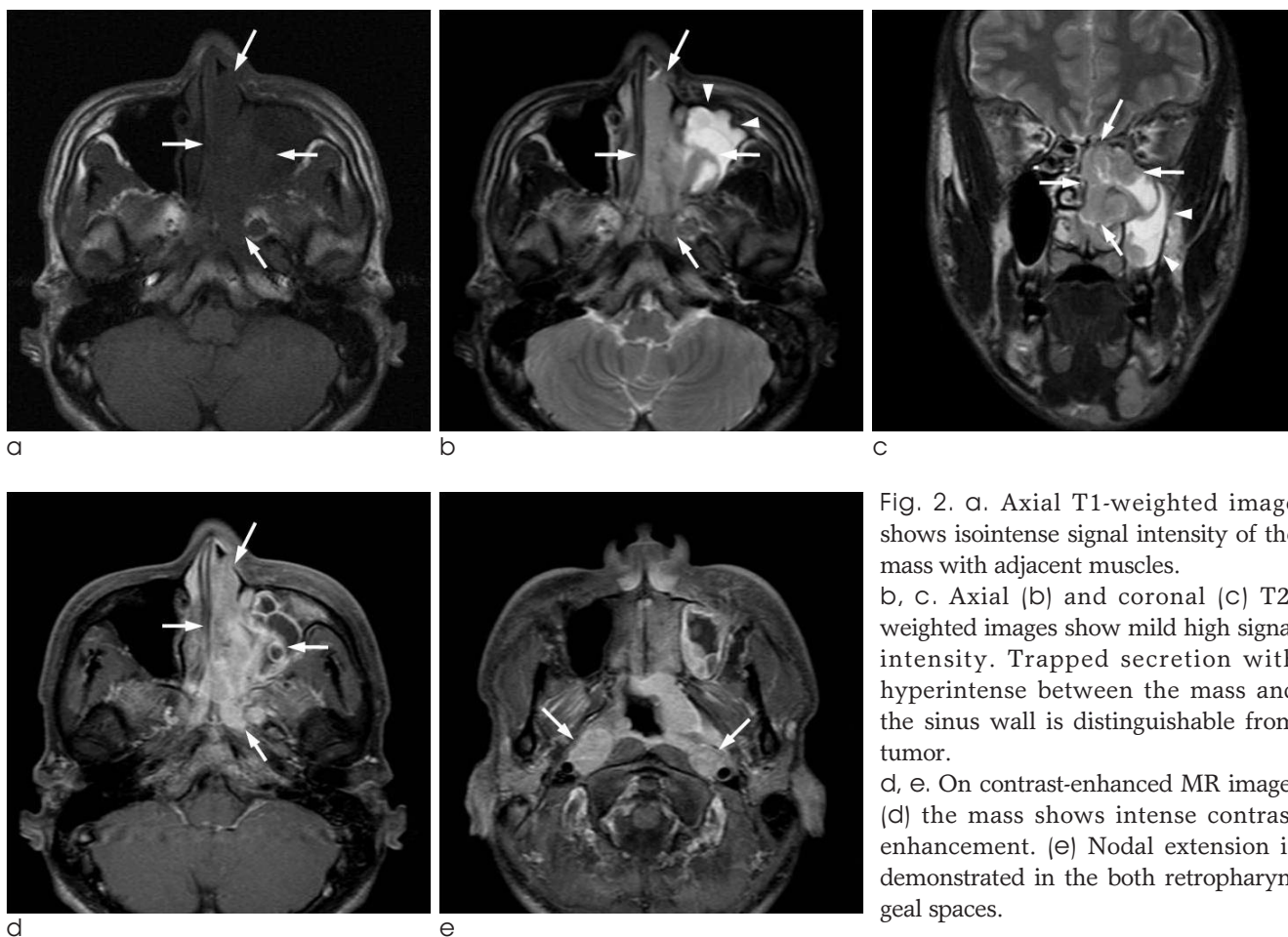


Fig. 2. a. Axial T1-weighted image shows isointense signal intensity of the mass with adjacent muscles. b, c. Axial (b) and coronal (c) T2-weighted images show mild high signal intensity. Trapped secretion with hyperintense between the mass and the sinus wall is distinguishable from tumor. d, e. On contrast-enhanced MR image, (d) the mass shows intense contrast enhancement. (e) Nodal extension is demonstrated in the both retropharyngeal spaces.

Biopsy was performed at the mass in the left nasal cavity. The histologic examination of the specimen revealed primarily undifferentiated tumor cells and scattered round and strap-shaped rhabdomyoblasts. Scattered tumor cells contain strongly positive cytoplasmic tails for desmin on immunohistochemical stain (Fig. 3). Thus the pathologic diagnosis was embryonal rhabdomyosarcoma.

Discussion

Adult rhabdomyosarcoma has several different characteristics compared to childhood rhabdomyosarcoma. First, the incidence of adult rhabdomyosarcoma is very low, especially in the embryonal type (2, 3). In contrast, in the pediatrics under 15 years, rhabdomyosarcoma is the most common mesenchymal malignancy. Second, the extremity is the most commonly affected in the adult rhabdomyosarcoma, however, the head and neck (especially the orbit) is the most frequent site in the childhood rhabdomyosarcoma. Lee et al. (3) reported only 1 case (2%) of nasal cavity rhabdomyosarcoma of the 51 adult rhabdomyosarcoma cases during a 5-year period. Third, overall survival is worse for adult than for children (2, 3).

In the present case, CT showed poorly defined, inhomogeneous soft-tissue mass destroying adjacent bones. After administration of contrast material, the masses enhanced generally to the same degree as adjacent mus-

cles. The MR images of the present case revealed a homogeneous mass, isointense or slightly hyperintense on T1-weighted images and slightly hyperintense on T2-weighted images, relative to adjacent muscles. After intravenous injection of contrast material, the mass enhanced more than adjacent muscles. These findings of CT and MR images are similar to the previous reports (3, 6, 7).

In the present case, it was difficult to differentiate trapped secretion and direct extension of tumor on CT scans because poor contrast resolution of soft tissues. However, trapped secretion between the mass and the sinus wall was distinguishable from tumor on MR images, especially on T2-weighted sequences; it was slightly hypointense on T1-weighted sequences and hyperintense on T2-weighted sequences and did not show enhancement. T2-weighted MR images and contrast enhanced T1-weighted images show better delineation of the tumor extent than on non-contrast enhanced T1-weighted sequences or CT scans. On T2 weighted and enhanced MR images, there were loss of normal dark signal of adjacent cortical bone and a hyperintense mass on the other side of the bone at posterior nasal septum, medial and posterolateral wall of antrum. This is the hallmark of bony destruction on MR images (3, 6, 7).

Rhabdomyosarcomas of the head and neck in adults are different from those of childhood in their site of origin, and the incidence and prognosis, however they

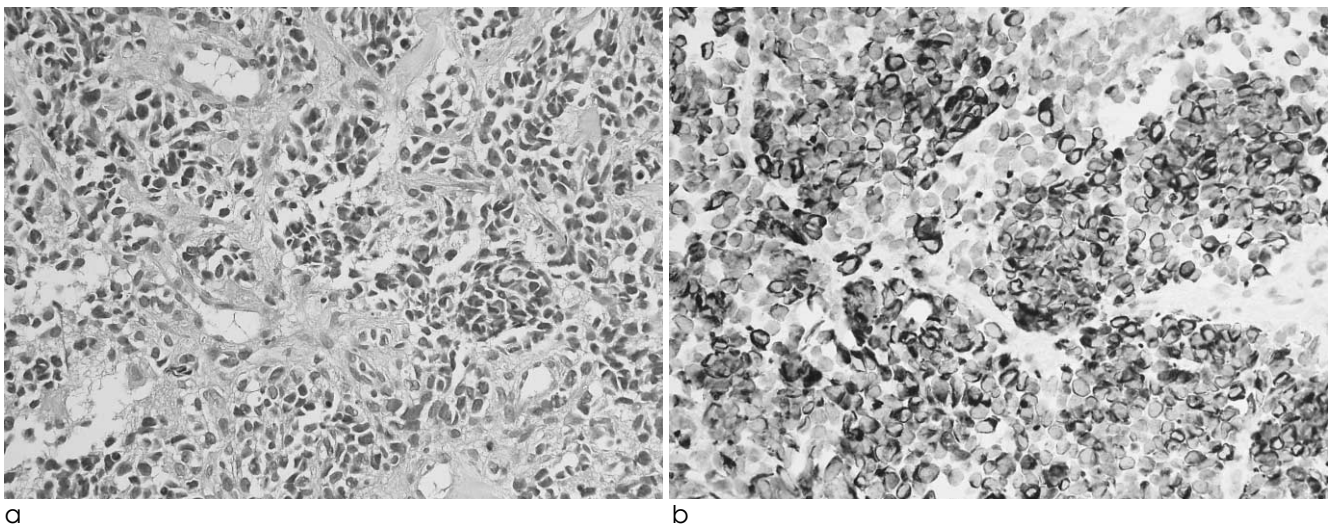


Fig. 3. a. Biopsy specimen shows primarily undifferentiated tumor cells and scattered round and strap-shaped rhabdomyoblasts (hematoxylin and eosin staining, magnification $\times 400$). b. Scattered tumor cells contain strongly positive cytoplasmic tails (Desmin).

show similar radiologic imaging findings. The differential diagnosis of rhabdomyosarcomas in the head and neck in adults includes the wide range of malignant tumors that may involve the head and neck, because of their similar radiologic imaging findings. In cases of multifocal lesions, one should consider lymphoma rather than rhabdomyosarcomas. And lymphoma is less often associated with invasion and destruction of adjacent bony structures. A fat component of liposarcomas and calcifications of chordoma, chondrosarcoma and osteosarcoma are helpful in the differential diagnosis. Other malignant tumors of the head and neck, such as squamous cell carcinomas, are usually found in older patients than rhabdomyosarcomas (3).

Conclusion

Embryonal rhabdomyosarcoma of the nasal cavity is very rare in adults. Rhabdomyosarcoma should be considered in the differential diagnosis of poorly defined soft tissue mass of the nasal cavity not only in children, but also in adults. In the patient with nasal rhabdomyosarcoma, the main strength of MRI over CT is the better delineation of tumor extent with differentiation from direct extension and trapped secretion because of superior contrast resolution of soft tissues, especially on T2-weighted images.

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성인의 비강에서 발생한 배아형문근육종: 증례 보고

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강민석 · 김학진

성인에서 두경부에 발생하는 배아형문근육종은 매우 드물다. 저자들은 좌측 비폐쇄, 동측 안구 및 협부통증과 다발성 경부결절을 주소로 내원한 24세 남자환자에서 발견된 비강내의 배아형문근육종 1예를 경험하였기에 문헌고찰과 함께 보고한다. 형문근육종은 소아에서뿐만 아니라 성인에서도 비강 내 종양의 감별진단에 포함되어야 한다.

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