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Alveolar Soft-part Sarcoma Metastatic to Palate : Report of 1 case

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Original Article

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Abstract

A case of metastatic alveolar soft-part sarcoma is presented and discussed. The lesion primarily arose from the left hip region and metastasized to numerous sites including the oral cavity. The review literature of this particular tumor is included in this article.

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Introduction

Alveolar soft-part sarcoma (ASPS) is a rare malignant neoplasm accounting less than one percent of all soft tissue sarcoma[1]. It was first named in 1952 by Christopherson et al[2]. Generally, it is a neoplasm of young adults but occasionally can be seen in neonates and in patients in their seventh decade. The incidence is higher in female than in male with a ratio of 2:1[3]. The average age of female patients was 20 years old while it was 10 years older in male patients[4,6].

APSP typically appears as a tender, slow growing mass usually painless, primarily located within deep soft tissue or skeletal muscle[2,7]. The anatomic distribution is interesting in that most of the cases arose in lower extrimities with 80% of those on the right side, especially the right thigh[6]. Since the lesions are usually asymptomatic, sometimes the primary tumor may not be detected. Anyway, some patients may develop malaise, weight loss, fatigue and fever[8].

The origin and histogenesis of ASPS have been studied by several investigators but still remains an enigma. The theories of non-chromaffin paraganglia[6,9-11] and pleuri-potential neural epithelial cells[12,13] have been proposed with little support. Several recent studies favored an origin from myogenic precursor[14-18] towards rhabdomyomatous differentiation[19,20]. Later, this theory has become more and more plausible. Anyhow, the conclusion of ASPS histogenesis have not yet been established and its theories of cellular origin are still controversial.

Pathologic findings

On gross examination, the primary tumor is usually well circumscribed and is frequently partially encapsulated but with invasion of the surrounding tissue. The tumor masses are firm and may be lobulated. The cut surface appears relatively uniform without characteristic features. It may be yellow or pink, showing homogeneous or stripped, honeycombed appearance. The size varies from 2-23 cm. in diameter[3,8].

Microscopically, the tumor is characterized by pseudoalveolar structures, namely alveolar nests, formed by groups of 4 to 50 cells delimited by delicate fibrovascular septa imparting and organoid pattern[8]. The individual cells are large, clear, either oval or polyhedral with large nuclei, prominent nucleoli and abundant acidophilic cytoplasm. The intra cytoplasmic rod-shaped crystalloid inclusions stain positively with PAS and are diastase resistant. Some diastase digestible PAS positive granules may also be demonstrated indicating the presence of glycogen within the tumor cells. Mitotic figures can be observed but not prominent. Invasion of the blood vessels by nests of tumor cells is frequent[1,9,10].

Several lesions bear a superficial histologic resemblance to ASPS, namely granular cell myoblastoma, alveolar rhabdomyosarcoma, paraganglioma, renal carcinoma (clear cell carcinoma), hepatoma, adrenal rest tumor and chemodectoma[3-5]. These tumors should be included in the differential diagnosis. The PAS stain is helpful in seperating ASPS from such tumors. In some instance, electron microscopic and immunochemical studies may be needed to make the definite diagnosis, especially in those cases that lack the typical crystals in the tumor cells[17,19,21].

Treatment and prognosis

ASPS grows slowly and usually stays indolent but the long term prognosis tends to be unfavorable. Metastatic rate is about 68%[22]. The patients often develop metastasis before the detection of primary lesion. Anyway, metastasis may occur as long as 15 to 28 years following resection of the primary lesion[1,22-24]. The common sites of metastases are lung, bone and brain. The 5 years survival rate ranges between 59-67%[6,22]. Survival decreased gradually with longer time follow-up. Neither tumor size nor site seemed to affect the prognosis since the patients eventually died of tumor even 28 years disease free interval.

With regard to the treatment of ASPS, wide local excision is the treatment of choice. The problem following excision is the high rate of local recurrence. Radiation has proved to give slightly better result when combined with surgery[6]. Chemotherapy has not yet give satisfactory result. ASPS should be always considered fatal because no case of life-time cure has been recorded with this tumor[6,8].

Head and neck primary ASPS is rare accounting about 25%[1] of overall cases with the orbit being the common locations. The tumors are found mostly in children[1,6,16]. ASPS of the oral region is unusual since only 12 cases have been reported during the past 33 years period[25]. Among these, base of tongue is an apparent area of predilection[1,2,25-28].

Case report

A 34-year-old black American male was well until October, 1980 when he noted a small, warm, hard tender swelling on the lateral aspect of left hip. He was treated with steroids but no any improvement and the mass was continued to grow slowly for 5 months

before the biopsy was done and reported as alveolar soft-part sarcoma. The patient underwent a wide excision of the tumor in March, 1981 and according to the pathological report, the margin of resection was clear. At that time, chest X-ray and CAT scan evaluation of this patient revealed pulmonary and brain metastases. He recieved chemotherapy and whole brain irradiation but failed to respond. The progression of brain metastasis caused blurred vision of the right eye. He also developed difficulty in breathing because of the lung metastasis. In October, 1981 he refused further chemotherapy for personal reason. The poor prognosis was discussed in January, 1982 and he was advised to report to the cancer center whenever he had any problem or discomfort. In March, 1982 he presented with a pedunculated lesion on the palatal marginal gingiva of teeth # 14 and 15. The mass was firm with sign of surface ulceration. It was measured $12 \times 12 \times 5$ mm. The lesion was locally removed for palliation.

The specimen recieved at the Oral Pathology Department, University of Maryland, was a formalin-fixed mucosal nodule. It was sectioned and submitted in toto for histopathological study.

Microscopically, the sections revealed ulceration with acute inflammation. The connective tissue was infiltrated by large ovoid cells with eosinophilic granular cytoplasm. The nuclei of the cells were hyperchromatic, pleomorphic and some mitoses were seen. These cells were arranged in small uniform, closely packed pseudoalveolar structures with delicate fibrous septa between them (Fig. 1,2). A microscopic diagnosis of "metastatic neoplasm consistent with alveolar soft-part sarcoma" was made. After the discussion of patient limited life expectancy, he made the decision that no heroic efforts be attempted any more. He lost follow-up and was expected of being succumbed to the disease soon after that.

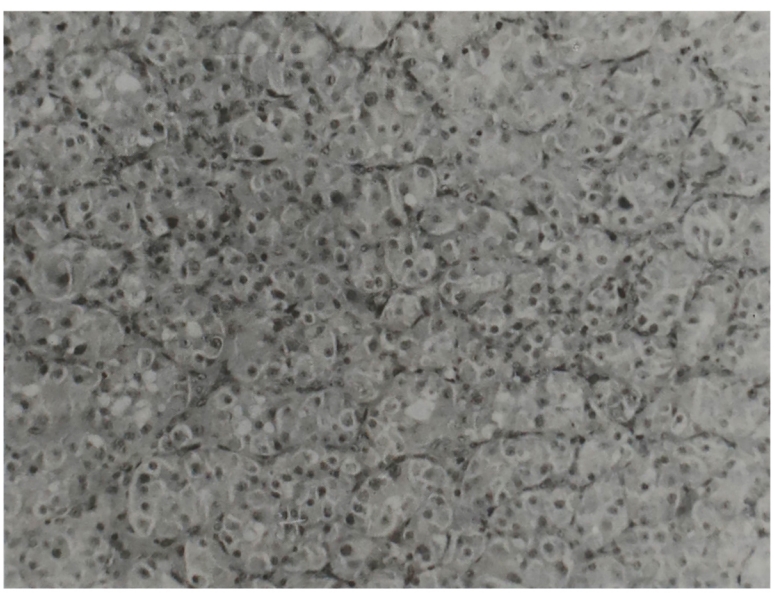

Figure 1 Tumor contains closely packed pseudoalveolar structures seperated by delicate fibrous septae. (H & E 50X)

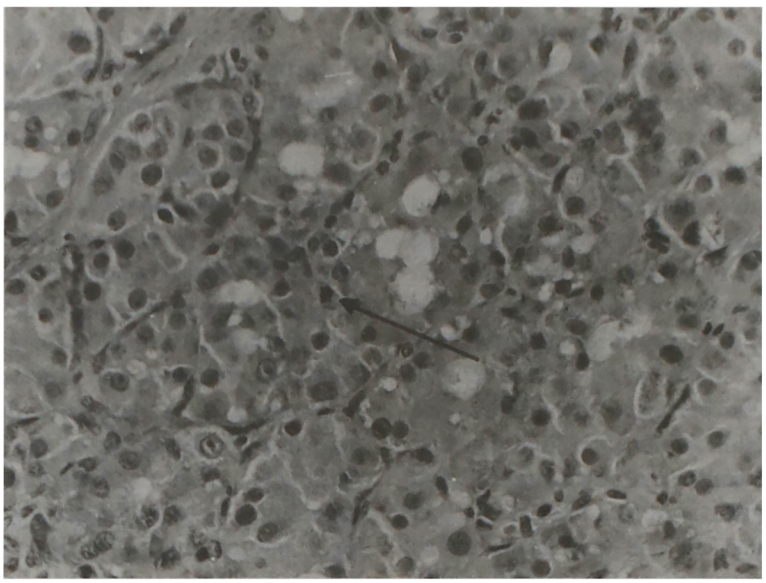

Figure 2 Large tumor cells exhibit eosinophilic granular cytoplasm and some mitotic activity. (H & E 100X)

Discussion

The case presented here is the first that revealed metastatic ASPS of the palate. Since ASPS is a rare malignant tumor showing tendency of rather late metastasis with the common secondary sites being lung, bone and brain, metastasizing to other soft tissue sites, especially to oral cavity, could be cosidered extremely rare. After the occurrence of this case, the following 6 years review of the literature showed no such metastasis.

Generally, any type of oral metastatic tumor is rare accounting for only 1 per cent of all malignant lesions found in oral cavity. Most of these lesions show no specific features. This usually raises the problem of diagnosis. The careful examination of the lesion accompanied with history taking and patient general health inspection would be helpful in making clinical judgement. Any ulcerated, rapidly growing, painful, easily bleeding, or presenting with neurological symptom such as numbness, need special attention and should be considered suspicious. Such lesions require promptly histological examination to rule out malignancy and give definite diagnosis. The histologic confirmation of the diagnosis of these lesions are important since it may be the first indication of undiscovered malignancy which might exist in a distant primary site. In known malignant tumor cases, the diagnosis of metastatic lesion helps to state the failure of treatment or ability to control the disease.

Unfortunately, based on data from the review of the literature, once the oral metastatic lesion was found, prognosis has always been poor. The patients eventually died from disseminated metastasis. This is true in most of the cases. Removal of these lesions is usually nothing more than palliative. Anyway, one should not indicate metastatic lesion as a hopeless case and reluctantly handle the patient. The value of definite diagnosis of such lesions should not be questioned since we could never only rely on clinical appearance of any case. Besides, the microscopic final diagnosis will serve as the basic data concerning therapeutic modality, prognosis, failure of the treatment and progression of the disease. So, in known cases of malignant tumor, it is strongly recommended that all the removed lesions be submitted for microscopic diagnosis.

Summary

ASPS is a rare malignant tumor of uncertain origin primarily located within soft tissue of the extremities. Its prognosis is poor despite of the sluggish growth because of the common local recurrence after treatment and metastases which can occur either early or very late after primary tumor resection with lung, bone and brain being the common secondary sites. Oral primary ASPS is rare but oral secondary ASPS is far more rarer. The oral metastatic tumor does not show any characteristic feature or symptom. Only histopathologic examination would help in making the diagnosis. Treatment of those lesions is local resection for palliative purpose. Overall prognosis

of the patients is grave because they usually end up with tumor recurrence and/or dissemination.

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บทวิทยาการ

รายงานผู้ป่วยอัลวีโอลาร์ ซอฟ-พาร์ท ซาร์โคมา ที่แพร่มายังเพดานปาก : รายงานผู้ป่วย 1 ราย

บทคัดย่อ

รายงานผู้ป่วยมะเร็งอัลวีโอลาร์ ซอฟ-พาร์ท ซาร์โคมารายหนึ่ง ซึ่งพบรอยโรคครั้งแรกที่ตะโพกข้าย และต่อมามีการแพร่กระจายไปหลายตำแหน่งรวมทั้งในช่องปากด้วย บทความนี้ได้รวบรวมข้อมูลในแง่ต่าง ๆ ของโรคนี้ที่เคยมีผู้รายงานไว้ด้วย

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