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Brain metastases revealing primary pulmonary soft part sarcoma

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Abstract

English:

Alveolar soft part sarcoma (ASPS) is a rare mesenchymal soft-tissue tumour. It commonly arises in the lower extremities in adults and the head and neck in children. Primary pulmonary involvement, without evidence of soft tissue tumour elsewhere, is very rare. We present the seventh case ever reported in the literature of primary ASPS of the lung, which was revealed by brain metastases in a 28-year-old male patient. The parietal brain tumour was resected. The radiological and histological study led to the diagnosis: metastasis of an alveolar sarcoma. The patient then had brain radiotherapy and chemotherapy. Using this case as a starting point, we reviewed the characteristics and the main therapeutic options in these uncommon neoplasms.

Keywords

Alveolar soft part sarcoma • brain tumour

Metastaze cerebrale care dezvăluie un diagnostic de sarcom pulmonar primar de părți moi

Rezumat

Romanian:

Sarcomul de părți moi alveolar este o tumoră rară, cu origine mezenchimală, care afectează țesuturile moi. La adulți, apare frecvent la nivelul extremității caudale, iar la copii la nivelul capului și al gâtului.Rareori tumora prezintă localizare unică, pulmonară, fară dovezi ale afectării țesuturilor moi. Lucrarea prezintă cazul unui pacient în vârstă de 28 de ani, diagnosticat cu sarcom de părți moi, cu punct de plecare pulmonar, prezența determinărilor secundare cerebrale fiind cea care a condus la formularea diagnosticului. Acesta ar fi cel de-al șaptelea astfel de caz raportat vreodată în literatura de specialitate. Tumora de la nivelul lobului parietal a fost rezecată, iar bilanțul imagistic și examenul histologic au confirmat diagnosticul de metastază cerebrală a unui sarcom alveolar. Pacientul a urmat ședințe de radioterapie și chimioterapie. Folosind acest caz ca punct de plecare, am revizuit caracteristicile și principalele opțiuni terapeutice în cazul acestui neoplasm mai puțin frecvent.

Cuvinte-cheie

sarcom de părți moi alveolar • tumoră cerebrală

Abbreviations

ASPS, alveolar soft part sarcoma; CT, computed tomography; MRI, magnetic resonance imaging

Introduction

Alveolar soft part sarcoma (ASPS) accounts for 0.5–1% of soft tissue sarcomas (1). In adults, it commonly affects the fascial planes or the skeletal muscles of the lower extremities, and in children, the head and neck region (2).

Reported cases of pulmonary ASPS are scarce. To the best of our knowledge, only six cases of pulmonary primary alveolar sarcoma were ever published. Herein we report the seventh case of pulmonary primary alveolar sarcoma revealed by multiple cerebral metastases.

Case report

A 28-year-old male patient, without medical history, was admitted to the Neurosurgery Department of Burns and

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Trauma Center, in Ben Arous, in January 2020 after experiencing persistent headaches during the prior 6 months and right hemibody weakness during the prior month.

The patient was conscious and had predominantly crural right hemiparesis. Otherwise, the clinical examination was normal. In particular, no respiratory symptoms were present: neither a history of breathlessness nor arterial blood gas or SpO_2 abnormalities were identified.

We performed a magnetic resonance imaging (MRI) scan, and it revealed two brain tumours, one in the left parietal lobe and the other in the occipital lobe. Both present a double-

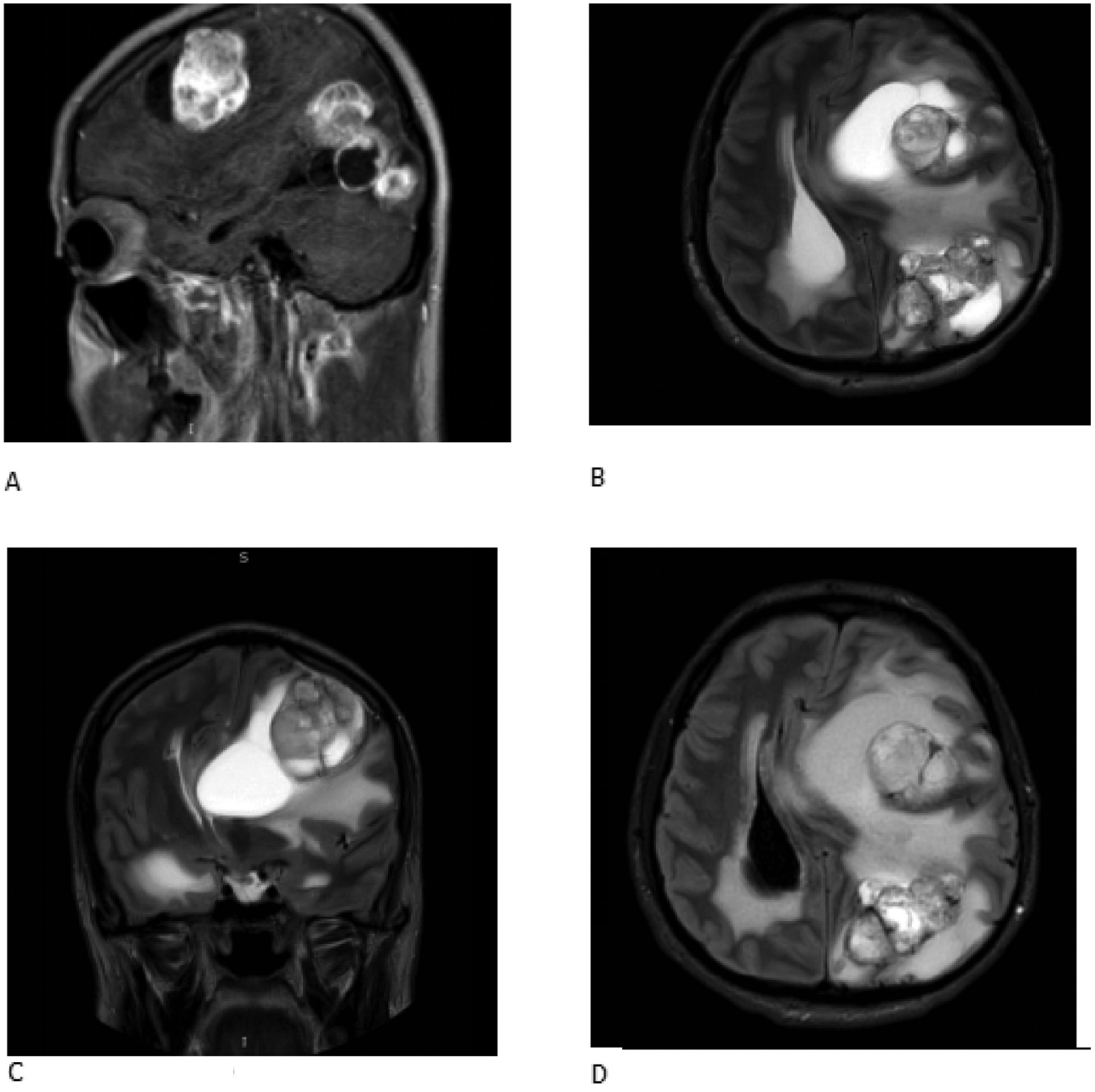


Figure 1. Sagittal section (A), Axial sections (B and D) and coronal section of a brain MRI on T1-WI with contrast injection (A), T2-WI (B and C) and T2-Flair (D) showing showing two left parietal and occipital tumours, with double-component, fleshy and cystic, hypointense on T1-WI, hyperintense on T2-WI. The fleshy part enhanced heterogeneously.

component, fleshy and cystic, with hyposignal T1, we propose this formulation: hypointense on T1-WI, hyperintense on T2-WI. The fleshy part enhanced heterogeneously (Figure 1).
The parietal lesion was causing oedema and a mass effect, making excision necessary. After craniometric spotting, a U-shaped incision was performed. The tumour was immediately accessible upon durotomy. The resection implied initial debulking within the tumour, followed by the careful dissection of its outlines.
The lesion was well-limited, firm and moderately hemorrhagic (Figure 2). Thus, no direct manipulation of the surrounding cerebral parenchyma was mandatory.
During anaesthesia, no respiratory issues occurred.
The postoperative period was uneventful. The patient did not present neurologic worsening. We performed a postoperative cerebral computed tomography (CT) (Figure 3) that showed neither complications nor enhancements suggestive for tumoural residue.
The histological examination described a tumoral proliferation, organised in solid nests and alveolar structures, indicating a metastasis of an alveolar sarcoma (Figure 4).
As part of the tumour-extension assessment, the patient underwent a thoraco-abdominopelvic CT-scan. It confirmed the existence of multiple nodules in both pulmonary fields and solid masses in the thoracic region. The nodules were round, with well-margined and smooth borders (Figure 4). We didn't find soft tissue tumours. We concluded that it was a primary pulmonary alveolar sarcoma revealed by brain metastases.
One month later, the patient underwent surgery during which we removed the occipital tumour. The headaches improved postoperatively. The patient had neither motor nor vision deficits.
We referred the patient for adjuvant treatment. He received targeted therapy-bevacizumab and radiotherapy.
After 6 months of follow-up, the scans were clear, with no recurrence of the brain metastases.

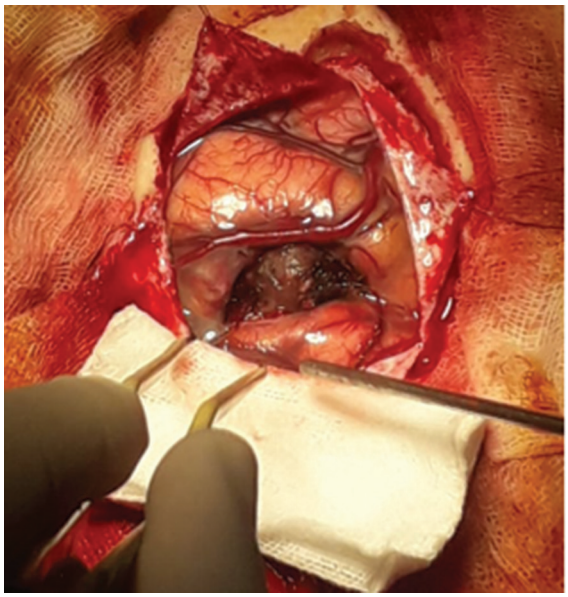


Figure 2. Intraoperative image showing the removal of the well-limited and firm lesion.

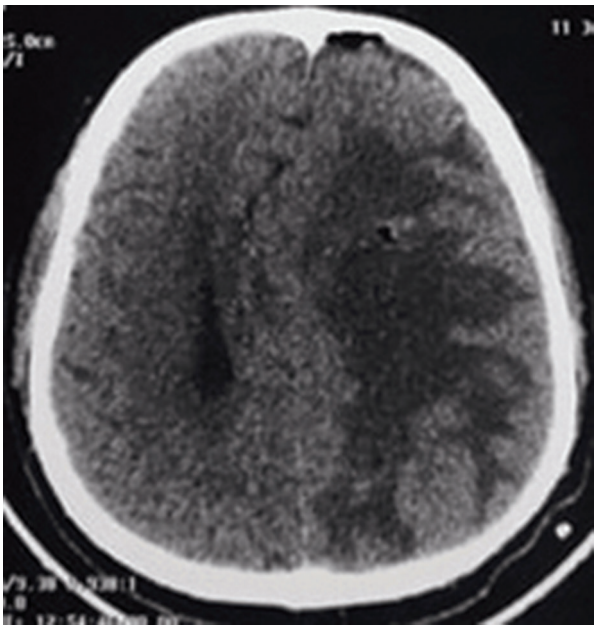
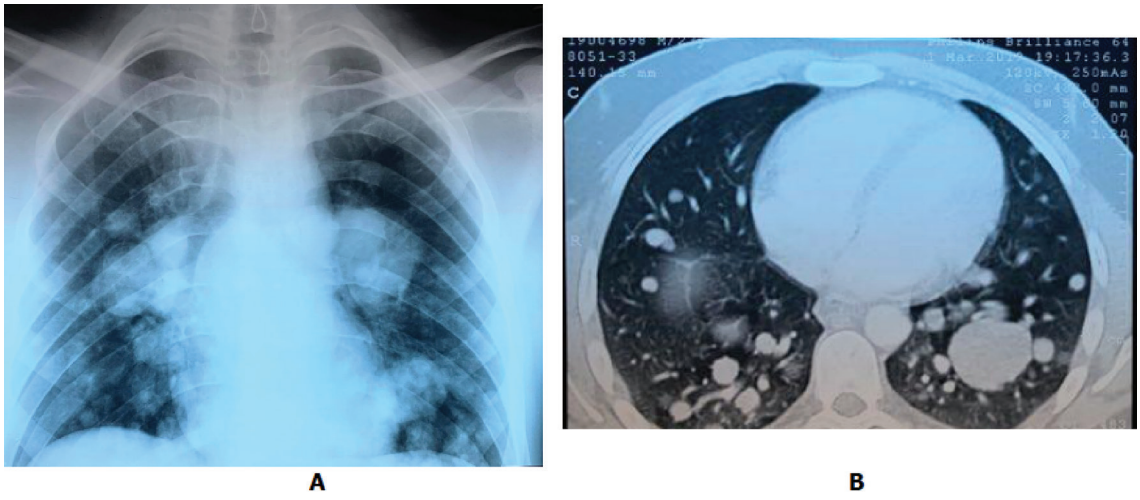


Figure 3. Postoperative CT-scan showing oedema. Neither hematoma nor tumour residue is noticed. CT, computed tomography.

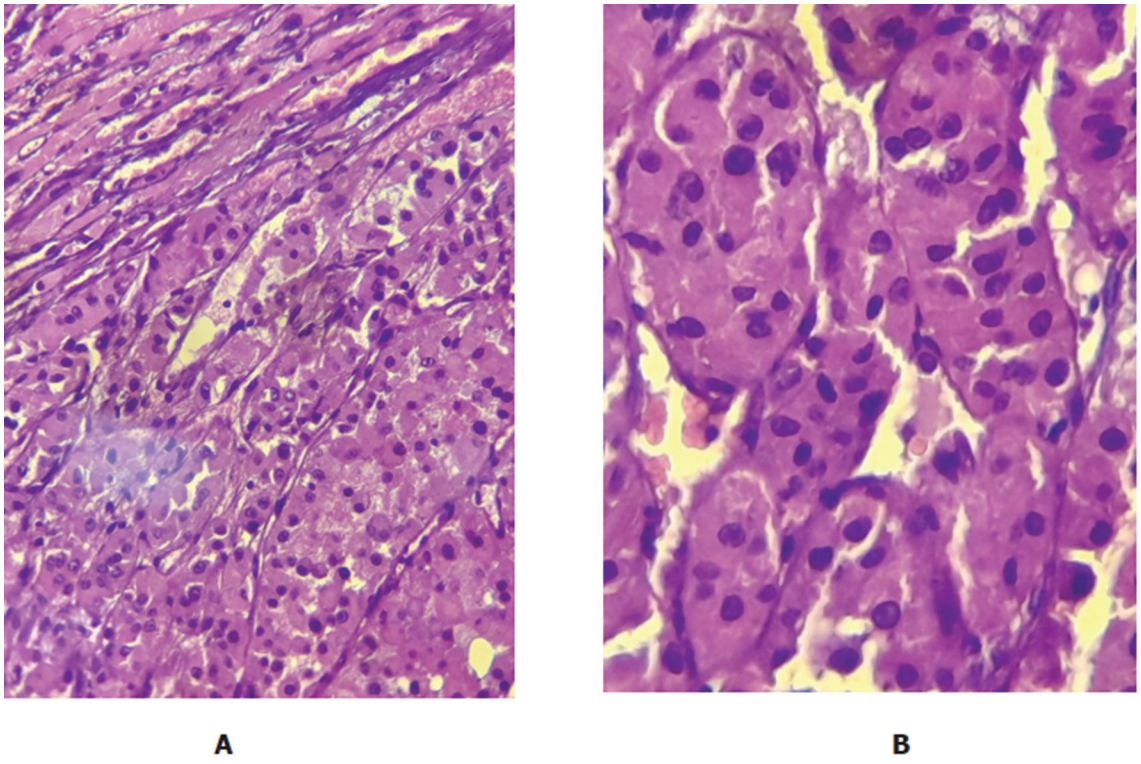
Discussion

ASPS occurs at all ages but is most prevalent in females aged 13–35 years (2). ASPS commonly presents as a slow-growing and painless mass, usually without functional impairment, but it is known to be highly malignant.
ASPS most often occurs in the extremities, followed by the head and neck region and trunk. ASPS can also involve the mediastinum, pituitary gland, stomach, spinal column, pulmonary vasculature, lung, breast and female genital tract. Tumours in the head and neck are most prevalent in children (3).
ASPS tumours frequently metastasise in the lungs, bones and brain.
The term alveolar refers not to a pulmonary origin but the appearance of irregular spaces within groups of cells when viewed through light microscopy (4). The tumour cells present some distinctive histologic features. They are rhomboid and present periodic acid Schiff (PAS)-positivity, diastase-resistance and rod-shaped crystals in a sheaflike arrangement in the cytoplasm.



A B

Figure 4. Thoracic X-ray (A) and CT-scan (B): showing multiple nodules and solid masses, rounded and smoothly contoured diffused by the two pulmonary fields. CT, computed tomography.



A B

Figure 5. Microscopical features of ASPS brain metastasis. (A) Hematoxylin Eosin ×10: alveolar structures and nests of tumour cells separated by fibrovascular septa and sinusoidal vascular spaces. (B) Hematoxylin Eosin ×40: large polygonal cells with distinct cell borders, granular cytoplasm, vesicular nuclei and prominent nucleoli. ASPS, alveolar soft part sarcoma.

Table 1. Clinical data of primary pulmonary ASPS in the literature.

Reference	Age	Sex	Revealing symptoms	Site of lung lesions	Therapy	Follow-up		Year
						Months	Results	
Tsutsumi and Deng (5)	42	Female	Hemoptysis	Right hilus	Surgery + chemotherapy	14	Died with disease Brain metastases	1991
Sonobe et al. (6)	24	Female	Chronic cough with mild chest pain	Right hilus (pulmonary vein)	Surgery	16	Alive with no evidence of disease	1994
Devisme et al. (7)	39	Female	Thoracic pain	Left lower lobe	Surgery	30	Lingual metastases Alive	1996
Trabelsi et al. (8)	49	Male	Thoracic pain	Right upper lobe	Chemotherapy	1	Died with disease	2007
Kim et al. (1)	42	Female	Incidental discovery	Right upper lobe	Surgery	25	Lumbar and occipital scalp metastases Alive	2007
Zhao et al. (9)	48	Female	Incidental discovery	Left hilus	Surgery	12	Alive with no evidence of disease	2015
Our study	28	Male	Persistent headaches revealing brain metastases	Bilateral nodules	Metastases surgery+ Brain radiotherapy + Chemotherapy	6	Alive	2020

ASPS, alveolar soft part sarcoma.

The origin of ASPS remains to be elucidated, although most researchers have hypothesised that it is myogenic (10).

Not infrequently, metastasis in the lung or other organs may be the first manifestation of the disease. Therefore, when the tumour is detected in visceral organs, other than soft tissues, it is mandatory to perform a thorough clinical and radiological evaluation before establishing the final diagnosis.

Primary ASPS of the lung is extraordinarily rare. To the best of our knowledge, our case is only the seventh to be reported. We presented a case of primary pulmonary ASPS revealed by brain metastases, without any evidence of soft tissue tumour elsewhere (Table 1).

Unlike in other published cases of pulmonary ASPS where the patient presented respiratory symptoms, in our case, the patient reported neurological symptoms (headaches).

The discovery of multiple bilateral nodules came as a surprise, as our patient had no respiratory symptoms. Moreover, the diffuse distribution of the pulmonary lesions contrasted with other published cases, where the pulmonary nodules were present unilaterally.

We were able to establish the ASPS diagnosis only after receiving the report of the histological examination of the brain lesion. In other cases, mainly because the patients presented pulmonary lesions, a fibrobronchoscopy was necessary to confirm the diagnosis.

No effective standard treatment is available for ASPS associated with brain metastasis (11). Therefore, patients with ASPS brain metastases are mainly treated via surgical resection to improve intracranial hypertension and other symptoms (12). In our case, we first did the resection of the parietal lesion and then of the occipital lesion. As ASPS tumours possess an abundant blood supply, en bloc rather than fractional resection should be performed. Surgical treatment followed by radiotherapy proved to prolong patient survival time (11, 13, 14). ASPS associated with brain metastasis is usually insensitive to chemotherapeutic drugs. At the moment, no specific chemotherapeutic treatment regimens are available.

According to previous prospective studies (15, 16), cediranib, a recently developed molecular-target drug, may effectively treat ASPS associated with brain metastasis with an overall remission rate of up to 35% and a 24-week control rate of 84%, thus maybe providing a novel clinical treatment option.

Conclusions

ASPS is a rare, malignant mesenchymal tumour. Cerebral metastases from lung primitive localisations are seldom. An aggressive therapeutic approach is commonly adopted to ensure the longest possible survival time. The optimum strategy associates total surgical resection, followed by adjuvant treatment: radiotherapy and chemotherapy. Molecular targeting drugs seem to offer new perspectives for optimised long-term remission. Our patient, a young man who presented with brain metastases related to a primitive alveolar sarcoma, is the perfect example of this therapeutic philosophy.

Future larger studies may provide possibilities for better standardisation of pre-existing protocols.

Authors contribution

KS and GG wrote the manuscript, AZ and MR performed bibliographic research, IZ and MB corrected the manuscript.

Conflict of interest statement

The authors declare declare having no conflicts of interest regarding this article

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