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Case Report / Приказ болесника

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Superior vena cava syndrome associated with alveolar soft part sarcoma – a possible paraneoplastic mechanism

Синдром горње шупље вене повезан са алвеоларним саркомом меког ткива – могућ паранеопластични механизам

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SUMMARY

Introduction Alveolar soft part sarcoma (ASPS) is an exceptionally rare soft tissue sarcoma, accounting for less than 1% of all cases. It typically presents as an indolent, slow-growing, painless mass. Superior vena cava (SVC) syndrome is usually caused by extraluminal compression from intrathoracic malignancies.

Case Outline We report the case of a 53-year-old male who developed acute symptoms of SVC thrombosis. Imaging revealed extensive venous thrombosis without significant extrinsic venous compression. The clinical findings mimicked classic SVC syndrome, yet the underlying mechanism was intraluminal obstruction caused by thrombus formation. Histopathological evaluation confirmed the diagnosis of ASPS.

Conclusion To our knowledge, this is the first reported case of ASPS presenting with SVC syndrome secondary to extensive thrombosis rather than compression. Although the underlying mechanism remains uncertain, this case raises the possibility of a paraneoplastic mechanism. Awareness of this atypical manifestation may facilitate earlier recognition and timely management in similar presentations.

Keywords: Alveolar soft part sarcoma; superior vena cava syndrome; thrombosis; paraneoplastic; chemotherapy

САЖЕТАК

Увод Алвеоларни сарком меког ткива (АСПС) је изузетно редак малигни тумор меког ткива, који чини мање од 1% свих саркома. Обично се манифестује спорим, индолентним растом и безболном масом. Синдром горње шупље вене (СВЦ) најчешће настаје услед екстралуминалне компресије интраторакалним малигнитетима.

Приказ болесника Приказујемо случај мушкарца старог 53 године који је развио акутне симптоме тромбозе горње шупље вене. Радиолошке методе откриле су опсежну венску тромбозу без знакова спољашње компресије. Клиничка слика је опонашала класични СВЦ синдром, али је основни механизам био интралуминална опструкција услед формирања тромба. Хистопатолошка анализа потврдила је дијагнозу АСПС.

Закључак Колико нам је познато, ово је први описани случај АСПС који се презентује СВЦ синдромом секундарним опсежној тромбози, а не компресији. Иако основни механизам остаје неизвестан, овај случај указује на могући паранеопластични механизам. Препознавање ове атипичне манифестације може допринети ранијој дијагностици и правовременом лечењу сличних стања.

Кључне речи: алвеоларни сарком меког ткива; синдром горње шупље вене; тромбоза; паранеопластички; хемиотерапија

INTRODUCTION

Alveolar soft part sarcoma (ASPS) is a rare subtype of soft tissue sarcoma, constituting less than 1% of all soft tissue sarcomas. It typically occurs in the lower extremities, head, and neck, affecting mostly adolescents and young adults aged 15–35 [1]. ASPS often metastasizes late, primarily to the lungs, bones, and brain. Despite its slow growth, prognosis is poor unless complete surgical resection is achieved. The standard treatment is wide surgical excision, and adjuvant radiotherapy is used if negative margins are not possible. Systemic chemotherapy has limited effectiveness in managing ASPS [1].

The aim of this report is to describe a rare presentation of ASPS manifesting with superior vena cava (SVC) secondary to extensive thrombosis and to discuss the possibility of an underlying paraneoplastic mechanism.

CASE REPORT

We present the case of a 53-year-old male who experienced sudden swelling of the face, neck, and upper limbs, along with dyspnea. He had no cough, hemoptysis, chest pain, or wheezing but reported erythema and pruritus on his forehead, cheeks, and upper chest (Figure 1). One week later, his symptoms progressed to dyspnea and dysphagia, with no fever, weight loss, or night sweats. Physical examination revealed suffusion of the right conjunctiva but no signs of icterus or clubbing. A firm, palpable right axillary lymph node was identified. The patient was diagnosed with SVCS and positioned with head elevation while receiving supplemental oxygen.

On admission, orthostatic changes were observed (supine BP: 130/80 mmHg, heart rate: 60–80 bpm; sitting BP: 85/55 mmHg, heart rate: 135 bpm). Oxygen saturation was 86% on room air, improving to over 95% with two liters of oxygen. Despite cyanosis and facial plethora (Figure 1), cardiovascular and respiratory examinations were otherwise unremarkable. Laboratory tests showed hemoglobin at 7.3 g/dL, platelets at 70 × 109/L, and white blood cells at 5.8 × 109/L. Ultrasonography revealed an extensive thrombus in the right jugular vein. Contrast-enhanced computed tomography of the chest confirmed a thrombus in the right brachiocephalic and internal jugular veins, and a six cm hematoma with ecchymosis within the sternocleidomastoid muscle (Figure 2). The patient was started on intravenous fluids and low-molecular-weight heparin. Due to ongoing bleeding, a core needle biopsy was performed, confirming sarcoma via immunohistochemistry (Figure 3).

Based on clinical presentation, imaging findings, and histopathological confirmation, a diagnosis of ASPS-associated SVCS secondary to extensive venous thrombosis was established. This presentation is highly atypical, as SVCS in malignancy most commonly results from extrinsic compression rather than intraluminal thrombosis. At the time of diagnosis, the patient was initiated on targeted systemic therapy for ASPS and continues on long-term anticoagulation.

Ethics: The patient agreed to have his photos and medical information published in the journal. He was aware that his name will not be revealed and that every effort will be made to maintain his anonymity.

DISCUSSION

ASPS is a rare soft tissue neoplasm characterized by historically uncertain histogenesis and an overall poor prognosis [1]. It predominantly affects females, usually involving the head and neck in children and the extremities in adults. Typically, tumor progression is slow, with symptoms developing over months to years; however, in our case, the presentation was acute and fatal. ASPS has a notable metastatic potential, commonly spreading to the lungs, bones, and brain [2]. Tumors in the extremities show metastases in approximately 20%-40% of cases at diagnosis [3]. ASPS is highly vascular, as reflected in its imaging findings. Hypervascularity with prominent draining veins and prolonged capillary staining is not exclusive to ASPS; it can also occur in hemangiomas or vascular malformations. In contrast to typical arteriovenous malformations, which demonstrate rapid contrast washout, ASPS is characterized by delayed contrast clearance and intense, persistent tumor enhancement. On MRI, ASPS typically presents a substantial solid component, whereas vascular malformations are composed entirely of vascular elements [2, 3]. However, this distinction was not clear in our case.

SVC syndrome affects an estimated 15,000 patients annually in the United States [4], with around 70% of cases linked to malignancies, while the remainder is mostly associated with medical devices. Accurate diagnosis is essential for selecting appropriate treatment, which may include radiotherapy, chemotherapy, surgery, or glucocorticoids, depending on the clinical scenario [4]. Among malignancies causing SVC syndrome, non-small cell lung cancer accounts for 50%, small cell lung cancer for 25%, and lymphomas for 10% [4]. Sarcomas, including ASPSs, are rare, with ASPS comprising about 1% of all soft tissue sarcomas [2]. Rare mesenchymal tumors of the thorax may present with unusual clinical syndromes and pose significant diagnostic challenges, as previously reported in rare intrapulmonary soft tissue tumors [5].

Paraneoplastic syndromes are distant effects of cancer, often unrelated to direct tumor invasion or obstruction, and typically manifest as neurological, hormonal, or hematologic symptoms. To our knowledge, this phenomenon has not been previously documented in ASPS. Cancer-associated hypercoagulability represents an important alternative explanation for the extensive venous thrombosis observed in our patient. Previous studies have demonstrated that tumor-associated thrombosis may result from multiple mechanisms, including tissue factor expression, thrombin generation, platelet activation, extracellular vesicles, and inflammatory cytokines, all of which contribute to a prothrombotic state [6]. However, the clinical presentation in our patient was unusual. ASPS is generally characterized by an indolent clinical course, whereas our patient presented with rapidly progressive SVCS as the initial manifestation of the disease in the absence of significant extrinsic venous compression. Although malignancy-associated thrombosis remains the most plausible explanation, the atypical presentation raises the possibility of a paraneoplastic mechanism. Nevertheless, in the absence of molecular, hormonal, or immunological evidence, a definite paraneoplastic syndrome cannot be established. The severity of these syndromes may not correlate with tumor

size and can precede cancer diagnosis. In our case, SVCS was the initial presentation, despite the tumor being less than four centimeters at diagnosis. Although curative surgery was not an option for this patient due to the advanced stage of ASPS, early suspicion of such syndromes may facilitate the detection of malignancy at an earlier stage.

Currently, the most commonly described paraneoplastic syndromes are associated with the secretion of functional peptides and hormones by the tumor, as seen in endocrine manifestations, or with immunological cross-reactions between normal host tissue and the tumor, as observed in neurological manifestations. In addition to these, venous thromboembolism (VTE) is also frequently reported in patients with malignancies. The incidence of VTE is relatively common in patients with sarcoma, reported in approximately 5–10% of cases [7]. Few studies have examined VTE in patients with sarcoma. For example, a SEER-Medicare retrospective study focused on older patients (over 65 years) reported a total VTE rate of approximately 16%, likely influenced by the advanced age of the cohort [8]. Another study reported a clinically relevant risk of venous thromboembolism in patients undergoing surgery for bone or soft-tissue sarcomas, particularly in the absence of thromboprophylaxis, with a non-statistically significant trend toward risk reduction when prophylaxis was used [9]. These findings, along with our results, indicate that VTE is not uncommon in patients with sarcoma and that both chemotherapy administration and the postoperative setting are significant risk factors.

CONCLUSION

This case highlights an unusual presentation of ASPS presenting with SVCS due to thrombosis, raising the possibility of a paraneoplastic mechanism. Awareness of such atypical thrombotic events may facilitate earlier diagnosis and improve timely management.

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Authors' contributions: All authors contributed equally to the conception, data collection, interpretation, and preparation of the manuscript. All authors critically reviewed and approved the final version of the paper and agree to be accountable for all aspects of the work.

Availability of data and materials: Data supporting the findings of this case report are available from the corresponding author upon reasonable request.

Conflict of interest: None to declare.

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Figure 1. Thrombosis-associated superior vena cava syndrome in a 53-year-old man; A – facial and neck plethora due to venous congestion; B – swelling of the right upper limb, a result of impaired venous return

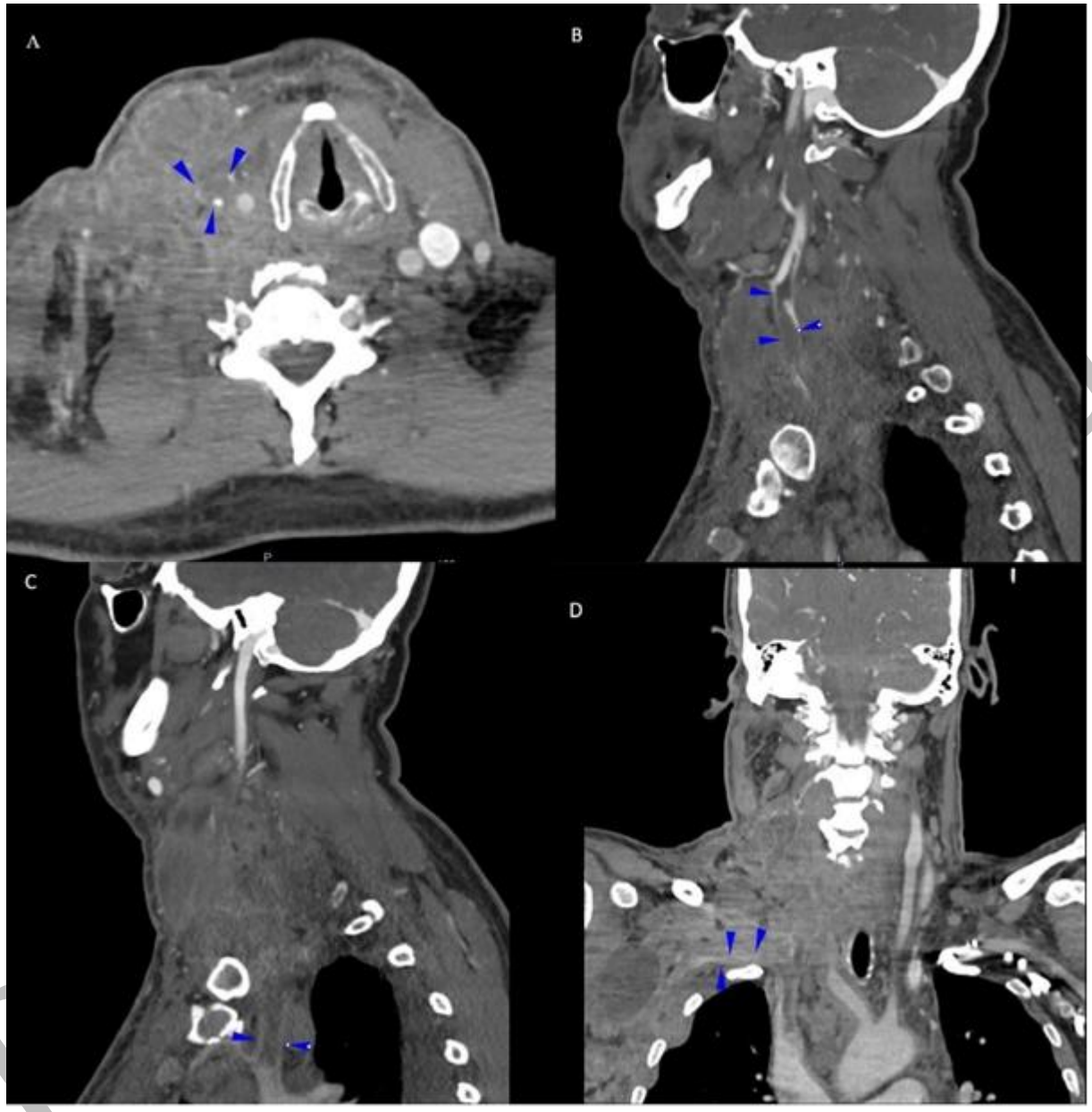

Figure 2. Computed tomography images; A – axial view shows right jugular vein obstruction by thrombus (arrow); B– sagittal view reveals jugular vein thrombus (arrow); C – sagittal view shows brachiocephalic vein thrombus (arrow); D – coronal view demonstrates mild extrinsic compression of the subclavian vein by the adjacent mass (arrow)

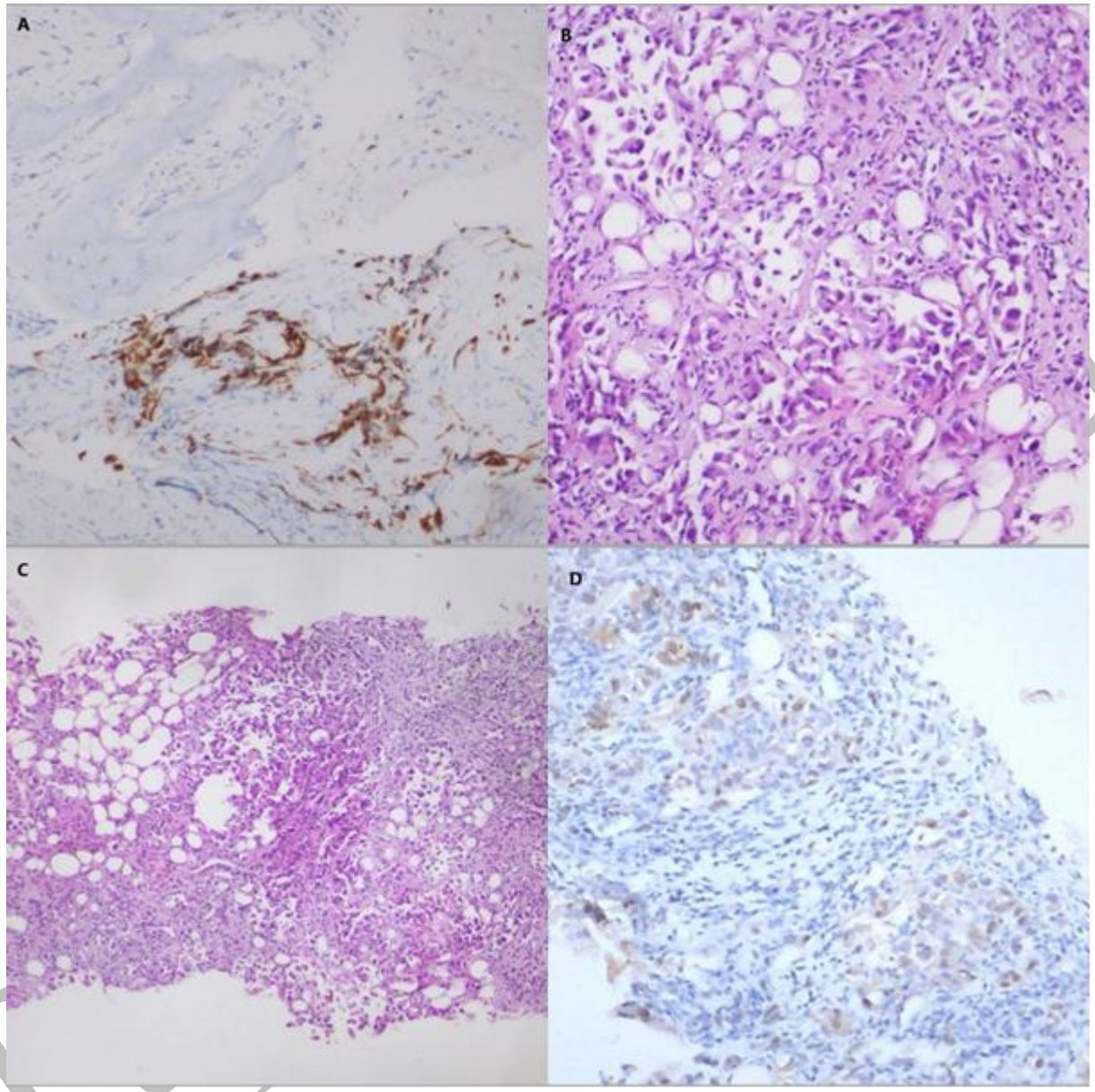

Figure 3. Pathology and histochemistry findings; A – cytokeratin-7 positivity in tumor cells near bone trabeculae (× 40); B – atypical tumor cells in an alveolar pattern in soft tissue (H&E, × 40); C – similar pattern in tru-cut biopsy (H&E, × 20); D – TFE3 positivity in tumor (× 40)

