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Scientific letter

Primary Pulmonary Ewing's Sarcoma: A Surprise Diagnosis in a 52-Year-Old Active Smoker

Sarcoma de Ewing pulmonar primario: una sorpresa diagnóstica en un fumador activo de 52 años

Dear Editor,

Ewing's sarcoma, also known as primitive neuroectodermal tumor (PNET), is the second most common bone cancer in children. It usually involves the pelvis and proximal long bones,[1] but in approximately 20% of patients, tumors are extraosseous and can arise in numerous organs. The most frequent extraosseous localizations include the chest wall, paravertebral and gluteus muscles, and the retroperitoneal space, but other rarer localizations have also been described.[2] Only sporadic cases of primary pulmonary Ewing's Sarcoma (PES) have been reported in the scientific literature, mostly in the form of case reports.

We report the case of a 52-year-old man who presented with a 3-month history of right-sided chest pain. The patient was an active smoker with a 40-pack-year history and no other significant medical history.

Physical examination was within normal limits. Chest X-ray showed homogeneous opacity with well-defined margins at the right apex of the lung. A computed tomography (CT) scan of the thorax revealed a large mass in the right upper lobe, measuring 24 cm × 13 cm with inhomogeneous enhancement on administration of contrast (Fig. 1A and B).

CT-guided biopsy revealed an undifferentiated tumor which consisted of large sheets of small round cells with perivascular arrangement (Fig. 1C) and an extensively necrotic area (Fig. 1D). Immunohistochemical staining was positive for vimentin, neuron specific enolase (NSE), synaptophysin, and CD99 (Fig. 1E).[3] The cells were negative for cytokeratin, TTF1, and CD 45. This profile suggested round cells of undifferentiated sarcoma from the Ewing family.

Radiological studies (whole-body CT) ruled out a primary tumor elsewhere, so a diagnosis of primary PES was made. The patient was referred to the multidisciplinary committee of a specialized center. Tumor size ruled out surgical treatment, and the decision was made to initiate neoadjuvant chemotherapy based on cyclophosphamide and vincristine. However, the patient died two months after the initial diagnosis.

The primary PES in a middle-aged active smoker is very unusual. Indeed, in our case, epithelial and neuroendocrine tumors of the lung would be the first diagnoses to be considered. Furthermore, the mean age at diagnosis of primary PES reported in the literature was 30.5 years, with only a few cases being diagnosed after the age of 50.[3]

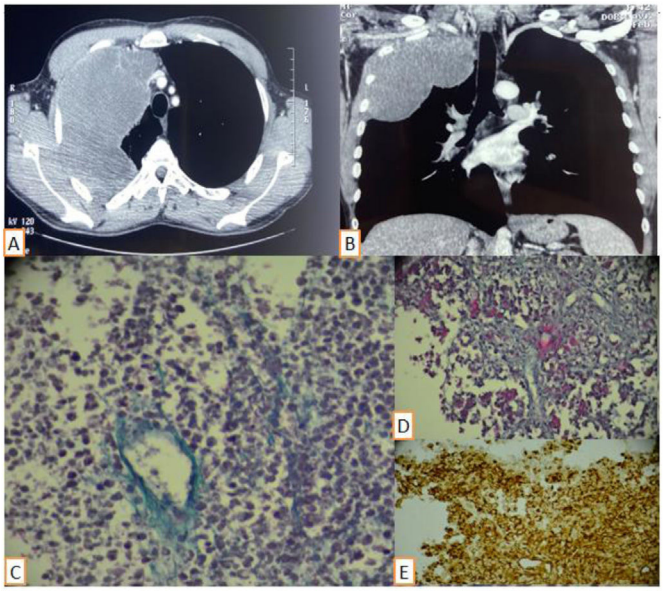

Fig. 1. (A and B) Computed tomography chest images showing a large, relatively well-defined, heterogenous pulmonary mass in the right upper lobe, measuring 24 cm × 13 cm. (C) Histopathology shows large sheets of small round cells with perivascular arrangement (HEA50 ×4). (D) Scattered pyknotic cells corresponding to apoptotic cells (HEA50 ×25). (E) Immunohistochemistry photomicrograph showing tumor cells positive for CD99.

Pathological and immunohistochemical analyses are essential for the diagnosis of PES and to eliminate other differential diagnosis. Strong staining for CD99 and neuroendocrine markers (synaptophysin, NSE, etc.) and negativity for cytokeratin, desmin and CD45 favor the diagnosis. Molecular testing is recommended for the study of EWSR1/FLI-1 fusions to ensure an accurate diagnosis.[4] In our case, however, due to limited laboratory facilities, immunohistochemical results were considered sufficient for the diagnosis and molecular analysis was not performed.

Generally, primary PES is an aggressive and lethal disease, especially when locally advanced and unresectable.[3] Tumor metastasis at diagnosis, large tumor size, and invasion of the heart are indicative of poor prognosis.[5]

In summary, despite its rarity, PES should be considered in the differential diagnosis of lung tumors, even in middle-aged active smokers. Early diagnosis is important, as it can enable radical surgical resection and improve prognosis.

Informed consent

Consent was obtained from a family member.

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Authors' contributions

All authors have contributed to all of the following:

- The conception and design of the study
- The drafting and revising of the article
- The final approval of the version

Conflicts of interests

The authors state that they have no conflict of interests.

References

1. Grünewald TGP, Cidre-Aranaz F, Surdez D, Tomazou EM, Alava E, Kovar H, et al. Ewing sarcoma. Nat Rev Dis Primers. 2018;4:5.
2. Lynch AD, Gani F, Meyer CF, Morris CD, Ahuja N, Johnston FM. Extraskeletal versus skeletal Ewing sarcoma in the adult population: controversies in care. Surg Oncol. 2018;27:373–9.
3. Fedeli MA, Marras V, Fara AM, Deiana A, Lobrano R, Cossu A, et al. Primary Ewing sarcoma of the lung: a systematic review of the recent literature. Ann Diagn Pathol. 2023;65:152152.
4. Brohl AS, Solomon DA, Chang W, Wang J, Song Y, Sindiri S, et al. The genomic landscape of the Ewing Sarcoma family of tumors reveals recurrent STAG2 mutation. PLoS Genet. 2014;10:e1004475.
5. Wang N, Dong SS, Wu CL, Wang Y, Meng L, Ren Y, et al. Rare primary pulmonary primitive neuroectodermal tumor: a case report and literature review. Onco Targets Ther. 2021;14:139–44.

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