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BRIEF REPORT

Extrapulmonary lymphangioleiomyomatosis in pelvic lymphadenectomy associated with invasive endometrial carcinoma

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KEYWORDS
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Abstract Lymphangioleiomyomatosis (LAM) is a rare disease occurring mostly in young women of childbearing age. This condition leads to a progressive cystic destruction of lungs due to the accumulation of epithelioid smooth muscle cells (LAM cells) in parenchymal and lymph nodes. LAM presents as a sporadic disease or a tuberous sclerosis complex (TSC) associated with other neoplasms within the family of the perivascular epithelioid cell tumours (''pecomas''). We present a case of a sporadic LAM affecting the lymph nodes of the pelvic chains observed as an incidental finding in a postmenopausal woman after a formal elective lymphadenectomy for endometrioid carcinoma.

PALABRAS CLAVE
Infangioleiomiomatosis;
Extrapulmonar;
Cáncer uterino;
Ganglios linfáticos pélvicos;
Inmunohistoquímica

Linfangioleiomiomatosis extrapulmonar en linfadenectomía pélvica asociada a carcinoma endometrial invasivo

Resumen La linfangioleiomiomatosis (LAM) es una rara enfermedad que afecta con más frecuencia a mujeres jóvenes en edad reproductiva y conlleva una destrucción quística progresiva de los pulmones por acumulación de células de músculo liso epitelioides (células LAM) a nivel parenquimatoso y en linfáticos. LAM se presenta como una enfermedad esporádica o integrante del complejo de esclerosis tuberosa (ET), asociado con otras neoplasias de la familia de tumores de células epitelioides perivasculares (''PEComas''). Presentamos un caso de LAM de tipo esporádico extrapulmonar en una mujer postmenopaúsica que afectaba a los ganglios linfáticos de las cadenas pélvicas con la peculiaridad de ser un hallazgo casual en la linfadenectomía electiva y reglada asociada a la cirugía de un carcinoma endometrioide.

Introduction

Lymphangioleiomyomatosis (LAM) is a rare disease that occurs almost exclusively in women of reproductive age.

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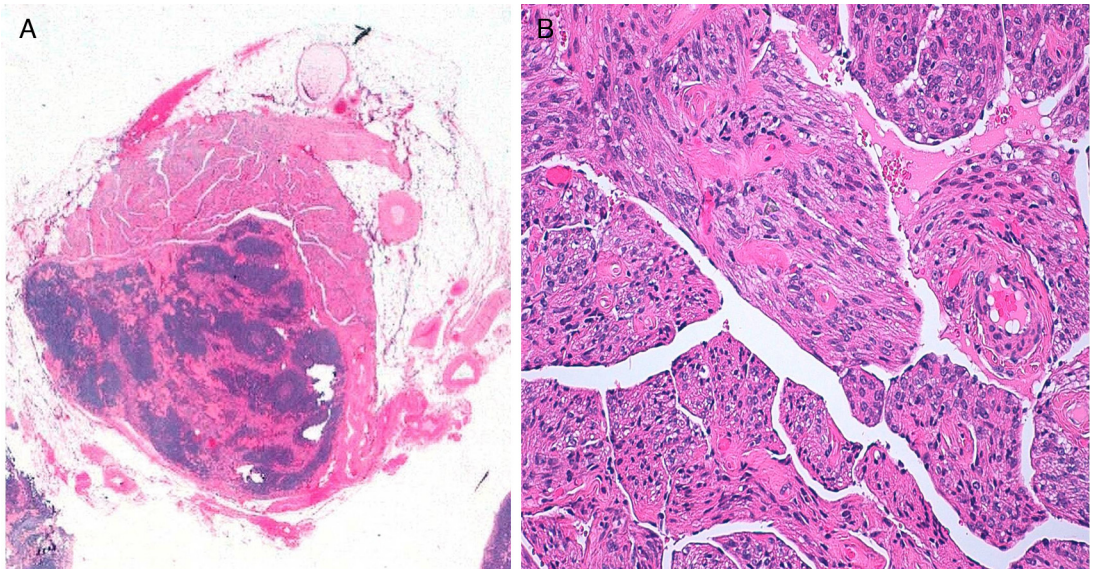

Figure 1 (A) Spindle cell proliferation arranged in nested pattern is seen in the periphery of the lymph node. (B) Spindle cell surround a network of channel-like spaces.

It is characterized by a proliferation of modified smooth muscle cells coexpressing melanocytic and muscle markers. As an entity, it is described stereotypically in the pulmonary tissue with extensive involvement, and it is considered a life-threatening disease. This condition can also originate in other organs such as lymph nodes, retroperitoneum and mediastinum. The extrapulmonary involvement is generally associated with lung disease, while the exclusive extrapulmonary LAM is highly unusual.[1,2] We present a case of extrapulmonary LAM found in pelvic lymph nodes associated with invasive endometrial adenocarcinoma.

Case description

A 72-year-old woman diagnosed with poorly differentiated endometrioid adenocarcinoma (more than 50% of the tumour was composed of solid masses of cells, with barely well-formed glands with marked nuclear atypia – grade 3 – and squamous differentiation) underwent surgery for scheduled total hysterectomy with double oophorectomy and bilateral pelvic lymphadenectomy in which a total of 34 lymph nodes were isolated. Microscopic examination revealed that fourteen showed a proliferation of epithelioid and spindle cells arranged in short fascicles and occasionally swirling (most of them under 3 mm in diameter). The cytological features were benign with inconspicuous nucleoli and clear or slightly eosinophilic cytoplasm. No cellular atypia or mitotic activity was observed (Fig. 1A and B). These cells surrounded a ramifying network of irregular channel-like spaces, partly lined by flattened cells. The structure of these channels showed a likely vascular or lymphatic origin. The architecture of the affected lymph nodes was preserved and the cell nests were located on the periphery without causing morphological distortion. No nodal metastasis of the primary tumour was observed. Immunohistochemistry revealed that both epithelioid and spindle proliferating cells showed intense expression for smooth muscle actin (AML), desmin (Fig. 2A and B); focally for HMB-45 (Fig. 2C) and MART-1/melan-A; microphthalmia transcription factor (MITF) (Fig. 2D); oestrogen and progesterone receptors (Fig. 2E) and intercellular channels were lined by spindle cells positive for D2-40 revealing lymphatic vascular nature (Fig. 2F).

Discussion

LAM is a very rare entity of unknown origin which almost exclusively affects young women of childbearing age with a prevalence of one case per million.[3] However, it is much more common in patients with TSC, where the symptoms of the disease are identified in up to 40% of adult women with this condition (TSC-associated LAM). Isolated cases of LAM in men and children associated with tuberous sclerosis have been described, but this is extremely rare. Seven similar sporadic cases in association with gynaecological lesions have been reported to date. Two of which were associated with an endometrioid adenocarcinoma in postmenopausal women and another was an incidental finding in the corresponding pelvic lymphadenectomy, as occurred in our case. These patients are often under going hormone replacement therapy with estrogens. A further case was associated wtih squamous cell carcinoma of the cervix and the remaining four with ganglionar endosalpingiosis.[2]

Regarding the reported cases of extrapulmonar LAM associated with gynaecological lesions, lung LAM was diagnosed six years after the retroperitoneal finding in one patient and coincidentally in a second case. In both patients, the lung LAM, as occurred in our patient, remained asymptomatic and did not require treatment.[2,4]

Tuberous sclerosis is an autosomal dominant neurocutaneous syndrome characterized by the presence of multiple hamartomas affecting mainly the central nervous system,