Cette tumeur est hautement agressive avec un potentiel métastatique important. Le traitement n'est pas encore codifié [5]. Il est habituellement multidisciplinaire associant une chirurgie, une radiothérapie et une chimiothérapie [4].

Conflit d'intérêt : aucun

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Alia Zehani, Ines Chelly, Haifa Nfoussi, Haifa Azzouz, Slim Haouet, Nidhameddine Kchir

Service d'anatomie pathologique. Hôpital La Rabta, Tunis 1007. Tunisie
Faculté de Médecine de Tunis
Université El Manar

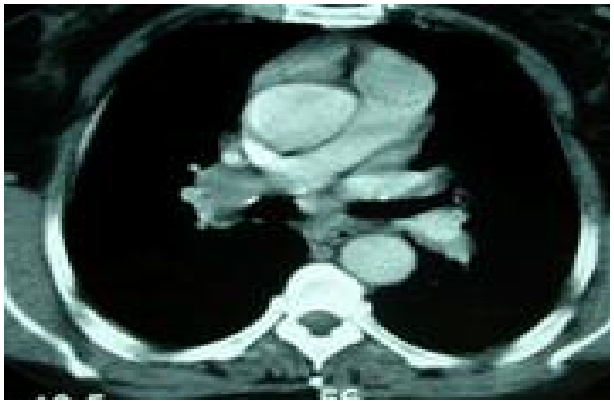
A case of anti-Jo1 myositis associated with sarcoidosis

Sarcoidosis is a systemic granulomatous disorder. It is rarely associated with other connective tissue disease. The association between systemic sarcoidosis and antisynthetase syndrome is extremely rare and is difficult to diagnose because of multiple common signs. Our aim is to report a new case of anti-JO1 myositis associated with sarcoidosis and to discuss the problems and difficulties to diagnose such association.

Case report

A woman of 63 years old visited our pulmonary outpatients department for dry cough and fatigue. She had dyspnea on exertion, weakness and myalgia of her arms and legs and polyarthralgia since few weeks. She had no previous respiratory problems. Physical examination revealed proximal muscle weakness. Laboratory tests showed elevated erythrocyte sedimentation rate to 105 mm. Serum creatine kinase level and lacticodesyhdrogenase were respectively elevated up to 3004 and 1200 IU/L. The tuberculin skin reaction was negative. Chest radiography revealed bilateral pulmonary interstitial lesions. Chest computed tomography showed bilateral ground glass lesions in the lower lung and bilateral hilar lymphadenopathies (fig: 1a, 1b). A pulmonary function test showed a severe restrictive syndrome. Bronchoalveolar lavage fluid showed increased total cell count with high proportion of lymphocytes (35%) and a high CD4 /CD8 ratio to 5.4. Transbronchial lung biopsy and salivary gland biopsy specimens showed non caseating granuloma formations. These findings suggested the diagnosis of systemic sarcoidosis but a connective tissue disease could not be excluded. This is why immunologic tests were realized; anti-JO1 was positive to 1/60. Histological examination of muscle specimen showed fragmentation of perimysial connective tissue and perifascicular myopathic changes compatible with myositis (fig.2). We conclude to the diagnosis of systemic sarcoidosis associated with antisynthetase syndrome. A high dose of corticosteroid (1mg /Kg/day of prednisone) was administrated for the rapid deterioration of respiratory function. Within 4 week corticosteroid therapy, fatigue, and polyarthralgia and CPK have decreased. With the same delay, functional parameters have been partially improved. Radiographic finding of interstitial syndrome were partially enhanced and mediastinal lymphadenitis have sub totally regressed within ten weeks. At follow up, muscle weakness remained although steroid therapy. Immunotherapy with methotrexate (20 mg /Week) was thereby administrated. The outcome was good. After four years follow up, the patient is now going well.

Figure 1a & 1b : Chest computed tomography showing bilateral ground glass lesions in the lower lung and bilateral hilar lymphadenopathies.



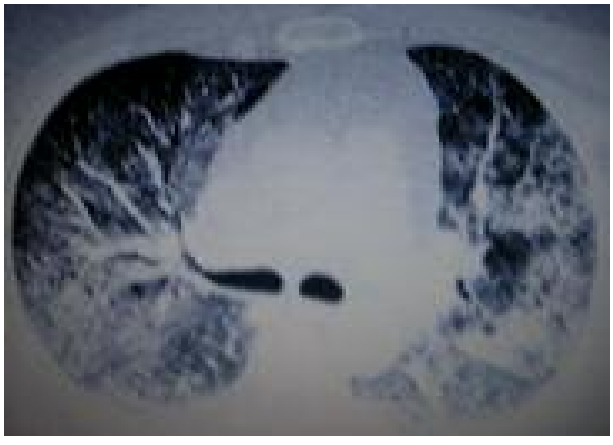
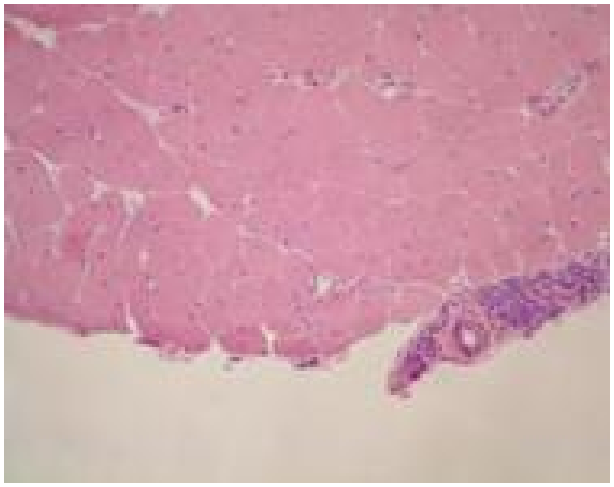


Figure 2 : Muscle biopsy specimen: fragmentation of perimysial connective tissue and perifascicular myopathic changes near connective tissue, compatible with myositis.



Conclusion

The association of antisynthetase syndrome and sarcoidosis is uncommon. To our knowledge, only one case of systemic sarcoidosis associated to antisynthetase syndrome has been reported in the English and French literature (1). The diagnosis of such association needs different investigations and multiple biopsies of the all affected organs. The characteristic feature of our patient is that multiple systems were affected in a mosaic pattern and biopsy specimens showed lesions consistent with both diseases.

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Sonia Toujani, Meriem Mjid, Bé chir Louzir, Yassine Ouahchi, Nozha Ben salah, Jalloul Daghfous, Nadia Mehiri, Jouda Cherif, Majed Beji.

Service de Pneumologie-Allergologie -Faculté de médecine de Tunis, Tunis El Manar Hôpital La Rabta - Bab Saadoun -1006 Tunis/ Tunisie

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Vascular reconstruction following an oncologic resection of a sarcoma of the groin: about two cases

Soft tissue sarcoma of the groin represents a real problem both for diagnosis and management. Its diagnosis is misleading especially in this location because it should be differentiated from common entities such as inguinal hernia or adenopathy. In the past the vascular structures invasion constitutes an indication for limb amputation but in recent years with the development of vascular reconstruction techniques, the limb sparing surgery becomes possible [1]. The 5-year survival rate ranges from 63 to 75% which demonstrates that the vessel involvement in itself does not represent a bad prognostic factor for overall and disease free survival when the tumor resection is wide enough [2].

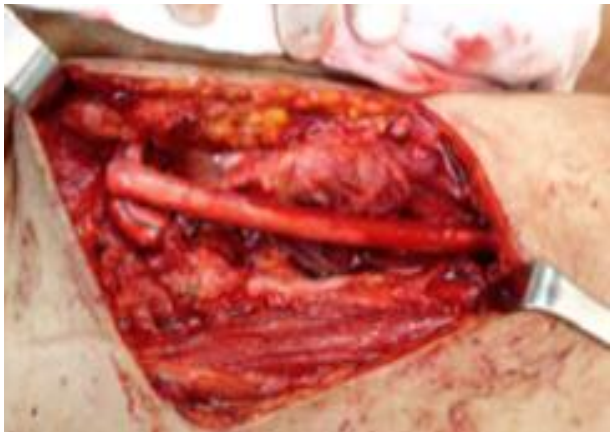
Schwarzbach recommended synthetic grafts for arterial and venous vascular substitute to reduce the operative time and to preserve the great saphenous vein (GSV) [3]. The venous reconstruction in case of groin sarcomas is unnecessary because of the establishment of collateral venous circulation due to the chronic tumoral compression.

We report the outcome of two cases of groin sarcomas that were completely resected with vascular reconstruction using synthetic grafts.

Case 1

A 57 year-old man had been referred in 2008 for a recurrence of an epithelioid hemangioendothelioma of the gluteal region two years after the initial treatment. We performed a reexcision of the tumor bed and an inguinal lymphadenectomy. The pathologic examination found a microscopic site of tumor with one positive lymph node. Adjuvant radiotherapy was delivered at the dose of 54 Gy. One year later, a second inguinal 30 mm recurrence was treated by wide excision. Eighteen months later, the patient presented a thrombophlebitis of the lower limb in which we discovered a deep third relapse. We performed a wide excision of the mass with arterial reconstruction using a PTFE graft. The patient is free of disease six months later.

Figure 1 : Figure 1: per operative view of the femoro-popliteal graft



Case 2

A 78 year-old man underwent in 1987 a resection of a sarcoma of the right thigh. On the postoperative course, the patient experienced an important hemorrhage that was controlled by ligation of the external iliac artery. The lower limb did not experience any acute ischemia event. He recently presented with a mass of the right groin that measured 70 mm and was attached to the deep plane. The CT scan showed a heterogeneous inguinal mass infiltrating the external femoral vessels. The biopsy concluded to leiomyosarcoma. A wide tumor resection through an inguinal incision was done including the femoral vessels. The restoration of the vascular