Pancreas Cancer-Associated Polymyositis of the Legs Regressing After Cephalic Duodenopancreatectomy: Case Report and Review of the Literature

Koceila Lamine Amroun¹, Louis de Mestier², Sophie Deguelte-Lardiere¹, Marie-Danièle Diebold³, Olivier Bouché², Reza Kianmanesh¹

Services of ¹General, Digestive and Endocrine Surgery, ²Hepatogastroenterology and Digestive Oncology and ³Anatomy and Pathological Cytology, "Robert-Debré" Hospital, Reims, France

ABSTRACT

Context Inflammatory myopathy, such as polymyositis, has been widely reported as paraneoplastic syndrome associated with various malignancies. However, its association with pancreas adenocarcinoma is very uncommon. Case report A case of a patient with paraneoplastic polymyositis of both legs associated with pancreatic adenocarcinoma is reported here. The diagnosis of polymyositis was highlighted by MRI and confirmed by histopathological examination. The surgical resection of the primary tumor led to the complete resolution of polymyositis with no further recurrence despite later metastases. Conclusion The association between pancreatic cancer and paraneoplastic polymyositis is very uncommon and has to be recognized by clinicians.

INTRODUCTION

The association between polymyositis and cancer has been widely reported [1, 2]. Although it may occur with most neoplasms, only a few authors have reported it as a pancreatic cancer-associated paraneoplastic syndrome [1, 3, 4]. The diagnosis of paraneoplastic myositis is usually suspected by clinical examination and standard imaging, and confirmed by pathological examination and clinical evolution under treatment. Data about its pathogenesis is scarce. A case of a patient with paraneoplastic polymyositis of both legs associated with a pancreatic adenocarcinoma is reported here. Thereafter, literature about its presentation and pathogenesis is reviewed.

CASE REPORT

A 47-year-old man presented with abdominal pain, jaundice, recent weight loss of 7 kg and asthenia. His past medical history included duodenal ulcer, renal lithiasis, appendectomy, alcohol abuse and smoking. There was no significant family history. His body mass index was 29 kg/m². On examination, there was jaundice and a diffuse abdominal tenderness without guarding. His laboratory investigation revealed the following: leukocytes 7,700 mm⁻³ (reference range: 4,000-10,000 mm⁻³), hemoglobin 99 g/L (reference range: 130-170 g/L), C-reactive protein 315 mg/L (reference range: 0-6 mg/L), gamma-glutamyltransferase 275 IU/L (reference range: 0-35 IU/L), alkaline phosphatase 276 IU/L (reference range: 32-120 IU/L), AST 100 IU/L (reference range: 10-30 IU/L), ALT 86 IU/L (reference range: 11-40 IU/L), total bilirubin 417 µmol/L (reference range: 4-17 µmol/L), conjugated bilirubin 309 µmol/L (reference range: 3-11 µmol/L), prothrombine rate 92% (reference range: 80-100%). A CT scan showed a 25 mm mass image located in the head of the pancreas, infiltrating the duodenum with a 2 cm portal trunk invasion. The superior mesenteric artery was clear. Biopsies were performed during endoscopic ultrasonography, and pathological analysis showed pancreatic adenocarcinoma. Despite an unsuccessful attempt of endoscopic biliary drainage, a biliary-digestive bypass was performed that led to effective biliary clearance. After multidisciplinary discussion, preoperative chemotherapy was decided, that consisted in four cycles of gemcitabine (1,000 mg/m², 1 injection per week during 3 of 4 weeks per cycle). The patient did not receive radiation therapy. During the fourth cycle of chemotherapy, the patient reported persistent pain of both lower limbs causing major difficulties in mobilizing. The pain was described as crushing sensation and cramps localized to the limbs. During the fourth cycle of chemotherapy, the patient reported persistent pain of both lower limbs causing major difficulties in mobilizing. The pain was described as crushing sensation and cramps localized to the limbs.
the anterior side of both thighs causing insomnia. Motor function, sensation and reflexes of lower limbs were normal. The skin had a paper-like texture in regard of the painful lesion. Biological markers were as follows: creatine kinase 1.6 N, lactate dehydrogenase 2.3 N and low levels of complement C3 and C4, with no evidence of inflammation. Doppler examination was unremarkable, hence excluding arterial or venous causes. MRI of lower limbs showed a diffuse T2 hyperintensity in both anterior muscles compartments associated with necrotic lesions (Figures 1 and 2), compatible with bilateral quadriceps polymyositis.

Skin and muscle were sampled. Pathological analysis of skin sample revealed scleroderma-like lesions without any immunohistochemical abnormality. Muscle examination revealed a global atrophy, large muscle necrosis with calcification and immune-histochemical overexpression of CD8+ T-cells (Figures 3 and 4).

After four cycles of chemotherapy, a CT-scan showed a 30% reduction of tumor size with no vascular invasion. The pain was intense and difficult to control despite of reducing and spacing gemcitabine doses and increasing opiates doses. Surgical resection was performed, consisting in a cephalic duodeno-pancreatectomy. There were no post-operative complications. Pathological examination of the resected specimen revealed a pT3N1aM0R0 infiltrating ductal adenocarcinoma (AJCC 2009). The complete resolution of muscular pain was remarkable, leading to a significant improvement of mobility and ability to walk three days after surgery. Levels of creatine kinase, lactate dehydrogenase, carcinoembryonic antigen and CA 19-9 became normal.

Six months later, two liver metastases were found on CT-scan and the CA 19-9 rate increased to 1,900 IU/L (reference range: 0-40 IU/L). Because of its initial efficacy, chemotherapy by gemcitabine was repeated at the complete dose during 5 months. No recurrence of polymyositis occurred. A revaluation CT-scan was performed afterwards and showed a moderate regression of the hepatic lesions. Nine months later, the patient presented with a community-acquired pneumonia causing severe sepsis leading to his death.

DISCUSSION

The association between cancer and inflammatory myopathy, such as polymyositis or dermatomyositis, has been widely reported but the pathogenesis remains elusive. This involves all histological types of malignancies. Adenocarcinoma, however, is the most frequently implicated, particularly from lung, ovarian, cervical, colorectal, gastric and breast origin [1, 2]. However, polymyositis has been scarcely reported as paraneoplastic syndrome associated with pancreatic adenocarcinoma [1, 3, 4]. Cancer may occur between

**Figure 1.** MRI (T1) axial section of both thighs with gadolinium injection. Necrotic aspect of both vastus lateral quadriceps.

**Figure 2.** MRI (T2 SPAIR) axial section of both thighs. Necrotic areas (hyperintensity) of both vastus lateral quadriceps.

**Figure 3.** Muscle sample with topographic coloration by hemalum, phloxine, safran (HPS). Fibrosis and atrophy of muscle fibers. Necrotic areas with ghostly muscle cells and calcifications.

**Figure 4.** Muscle sample with immunohistochemical preparation. CD8+ T-cells in contact with altered muscle cells.
A case of paraneoplastic polymyositis associated with pancreas adenocarcinoma is reported here. Muscular disorders regressed quickly after the surgical removal of primary tumor, strengthening the hypothesis about the cancer-related origin of these symptoms. Polymyositis is a well-described paraneoplastic syndrome, but its association with pancreas cancer is very uncommon and has to be recognized by clinicians.

**Conflicts of interest** All authors declare no conflicts of interest.

**References**