Sjogren syndrome and abdominal adhesions. Could they be related?

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ABSTRACT

Background: Sjögren syndrome is a chronic systemic inflammatory disorder (classified as an autoimmune disorder) characterized by lymphocytic infiltrates in exocrine organs. Complications related to Sjögren syndrome include SLE (systemic lupus erythematosus) and RA (rheymatoid arthritis) , infection of the parotid gland, typically staphylococcal, streptococcal, or pneumococcal, the appearance of parotid tumors, fetal loss during pregnancy in antiRo/SS-A-positive women and the occurrence of pseudolymphomas and non-Hodgkin B-cell lymphomas. Here we describe intraabdominal adhesions found in a patient with known Sjögren syndrome and no other known health problems.

Case report: We report the case of a 50-year-old menopausal woman with known Sjögren syndrome. She did not have any other medical issues. Most importantly, she did not report any prior abdominal surgery. The patient suffered from ovarian cysts and was scheduled to undergo elective laparoscopic ovary removal. Intraoperatively, the patient was found to have thick and extensive adhesions between the omentum and the abdominal peritoneum.

Conclusion: This case led us to wonder whether there might be a connection between Sjögren syndrome and the occurrence of adhesions. Further research is needed in cases with the same presentation. If we were able to clarify a common pathophysiology that connects the occurrence of intraabdominal adhesions with the syndrome, the disease itself might be better understood.

KEYWORDS

Sjögren syndrome; adhesions; laparoscopy; scleroderma; symphysiolysis; case report.

Core tip: *To our knowledge, Sjögren syndrome has never been associated with intrabdominal adhesions. This case report describes the occurrence of intrabdominal adhesions in a patient with known Sjögren syndrome and no other factors that might participate in their occurrence. The patient had ovarian cysts, which it was decided to remove laparoscopically. During the procedure extensive and thick adhesions between the omentum and the abdominal peritoneum were discovered.*

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Introduction

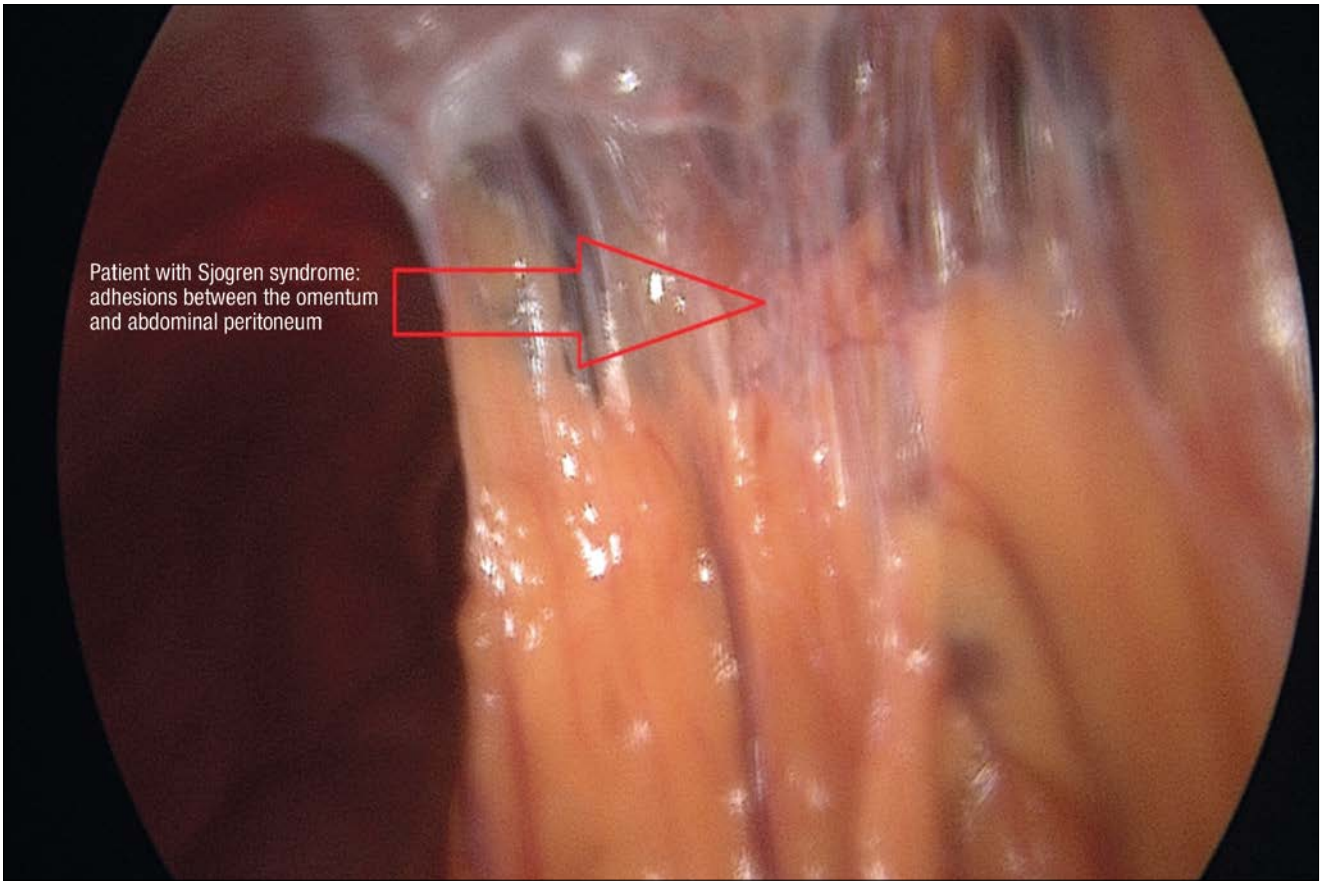
Sjögren syndrome is a chronic systemic inflammatory disorder (classified as an autoimmune disorder) characterized by lymphocytic infiltrates in exocrine organs. The disorder typically develops gradually beginning in middle adulthood, but it can occur at any age and mostly affects women (fifty to sixty years old). It can be associated with other autoimmune diseases (scleroderma, systemic sclerosis, cryobulinemia, polyarteritis nodosa) [1].

Intra-abdominal adhesions may be congenital or acquired. Congenital adhesions arise during physiological organogenesis or can be traced back to abnormal embryonal development of the abdominal cavity. They are usually asymptomatic, not extensive and are diagnosed incidentally [2]. We present the case of a 50-year-old menopausal woman with known Sjögren syndrome, who had thick and extensive adhesions between the omentum and the abdominal peritoneum, which were found during laparoscopic surgery for adnexectomy. Since these could not be postoperative complications, given that she had no history of previous surgeries, the hypothesis arose that they might be attributed to her autoimmune disease.

Case presentation

The 50-year-old woman with a history of Sjögren syndrome was postmenopausal with no other medical issues. She was a non-smoker, and had normal vaginal swabs (no chlamydia or other vaginal infection) and normal cervical cytology. She had had one vaginal delivery and most importantly, as mentioned before, she reported no prior abdominal surgery and had no past medical history indicative of any kind of abdominal inflammation. On an abdominal ultrasound, for vague abdominal discomfort, she was incidentally found to have bilateral

Figure 1 Intraoperative laparoscopy photo of adhesions. Thick and extensive adhesions between the omentum and the abdominal peritoneum were observed.



benign-looking ovarian cysts. Her blood serum CA125 levels were normal. The risk malignancy index was 54 and the IOTA classification was 98.4% for a benign tumor. Due to the size of the cysts, which each measured more than 6 cm, and her clinical presentation, an elective laparoscopic adnexectomy was offered, to which she consented.

Intraoperatively, we found thick and extensive adhesions between the omentum and the peritoneum covering almost all the abdominal cavity. We carefully removed the adhesions one by one using bipolar diathermy and cold scissors, until the omentum was completely released. The procedure was completed with the bilateral adnexectomy and removal of the specimens with the use of an endobag. Pathology was negative for malignancy, and cystadenoma was diagnosed on both ovaries.

Discussion

In most cases, the main symptom of Sjögren syndrome is xerophthalmia (dry eyes), xerostomia (dry mouth), and parotid gland enlargement. Other extra-glandular features are arthralgia, arthritis, myalgia, anemia, neuropathy, renal tubular acidosis, Raynaud phenomenon, pulmonary disease, leucopenia, lymphadenopathy, vasculitis (which can cause a rash that looks like small bruises or reddish-purple spots), and lymphoma. Complications related to Sjögren syndrome include SLE and RA, infection of the parotid gland, typically staphylococcal, streptococcal, or pneumococcal, parotid tumors, fetal loss during pregnancy (in antiRo/SS-A-positive women), pseudolymphomas, and non-Hodgkin B-cell lymphomas [3,4].

The etiology of the syndrome is not entirely known, but it seems to be due to an ongoing interaction between the innate and acquired immune systems. HLA-DR52 occurs in primary Sjögren syndrome with a frequency of about 87%, but genetic associations differ between ethnic groups; the syndrome can possibly be triggered by certain viruses, such as HIV, HCV, CMV, EBV, HHV-6. Damage and/or cell death due to viral infection or other causes may provide triggering antigens to Toll-like receptors in or on dendritic or epithelial cells. These cells, recognizing pathogen-associated patterns, are activated and begin producing cytokines, chemokines, and adhesion molecules. As T and B lymphocytes migrate into the gland, they themselves become activated by dendritic and epithelial cells, thereafter acting as antigen-presenting cells [1,3].

Peritoneal adhesions can be acquired or congenital. Congenital adhesions are formed during organogenesis (for example the attachment of the sigmoid colon to the left pelvic wall), or can be traced back to abnormal embryonal development of the abdominal cavity, and are not usually extensive. They can be asymptomatic and often are diagnosed incidentally. Postmortem examination of patients who had not undergone surgery identified post-inflammatory adhesions in 28% of cases.

These are caused by intra-abdominal inflammation or can be attributed to peritonitis, endometriosis, radiotherapy and long-term peritoneal dialysis. Postoperative adhesions usually form after 50% to 100% of all abdominopelvic interventions. They develop as a result of wound healing and are influenced by various factors [2].

This case report led us to wonder whether Sjögren syndrome and the occurrence of intrabdominal dense adhesions might be connected. A search of the PubMed and Scopus databases for relevant literature revealed no reports of similar cases. We searched the databases using the entries "Sjögren disease and adhesions" and "adhesions due to autoimmune diseases" and found no relevant studies. Also, detailed consultation of classical textbooks revealed no description of any connection between the two medical entities [4,5].

On the other hand, autoimmune diseases such as scleroderma have been reported to display complications, which include adhesions on the skin and internal organs, resulting from a pathogenetic mechanism that has not been fully clarified. There is a hypothesis that collagens I, IV and, VI, fibronectin and laminin participate in the formation of the adhesions [6]. As it is known that Sjögren syndrome is associated with scleroderma and other autoimmune diseases [7], it is reasonable to assume that Sjögren syndrome could trigger the formation of abdominal adhesions either directly or indirectly. The pathogenetic mechanism could be investigated on an autoimmune basis.

Conclusion

This case led us to wonder whether Sjögren syndrome and the occurrence of adhesions, like the ones we encountered, might be connected. We believe that there is a need for further investigation of the pathophysiology and the pathogenetic pathway that might connect the occurrence of intraabdominal adhesions with the Sjögren syndrome, either directly or indirectly. Also, we believe that publication of such cases might motivate more medical professionals to look into this matter in more depth.

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