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Diagnosis of IgG4-related systemic disease by cytology of large pericardial effusion with fine needle aspiration

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3 **Diagnosis of IgG4-related systemic disease by cytology of large**
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6 **pericardial effusion with fine needle aspiration**
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48 **Key words:** IgG4-related disease; Pericardial effusion; Fine needle aspiration cytology;
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51 Retroperitoneal fibrosis
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3 IgG4-related disease has characteristic features as follows; Serum IgG4 is
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6 prominently elevated, IgG4-positive plasma cells infiltrate in involved tissues and
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9 various mass-forming lesions with fibrosis, such as the autoimmune pancreatitis,
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12 salivary gland and retroperitoneum. As this disease comes to attention, the reports of
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15 IgG4-related disease have been increasing, but the pathogenesis and exact frequency of
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18 the disease remains unknown.
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22 We present the case of a 69 year-old male who was admitted to our hospital for
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25 evaluation of lower legs edema in July, 2010. He first noticed lower legs edema in
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28 2009 and visited a local clinic, and was diagnosed deep vein thrombosis, received
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31 anti-coagulation therapy. However, his edema was getting worse and hydrocele testis
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34 was appeared. When he visited our office, he complained of trouble in walking with
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37 lower legs edema. On physical examination, his body temperature was 36.2°C, blood
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40 pressure 152/82 mmHg, and pulse rate 60 per minute. There was no murmur in his
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43 cardiac sounds and lungs were clear. On abdominal examination, it was slightly
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46 swelling but not any mass were noted. His trunk was edematous, especially in lower
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49 legs. The thyroid gland, salivary glands and lymph nodes were not palpable. He had
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52 never experienced dry eye and dry mouth. Laboratory findings were as follows: WBC
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55 4390/ μ L, hemoglobin 10.6 g/dL, platelet 27.3×10^4 / μ L, BUN 10 mg/dL, creatinine 1.04
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3 mg/dL, BNP 119.0 pg/ml, CRP <0.10 µg/L. Liver function test was normal.
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6 Urinalysis did not demonstrate any protein on dipstick. Rheumatoid factor,
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9 anti-nuclear antibody, anti SS-A and anti SS-B, antibodies and antineutrophil
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12 cytoplasmic antibody were all negative. IgG, IgA, IgM and IgE were 1769.0 mg/dL,
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15 272.7 mg/dL, 74.8 mg/dL and 276 mg/dL, respectively. Serum level of IgG4 was
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18 elevated by 408 mg/dL. A chest radiograph revealed marked cardiomegaly.
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22 Echocardiography revealed marked pleural effusion with slightly collapsed right atrium.
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25 Computed tomography (CT) revealed also large pericardial effusion (Fig 1A) and low
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28 density area around aorta (Fig 1B). The mass, which was suspected retroperitoneal
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31 fibrosis (RPF), compressed inferior vena cava and right urinary duct, and induced lower
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34 legs edema and right hydronephrosis. After admission, a ureteral stent was placed in
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37 order to treat the hydronephrosis. Lip biopsy was done, but there was no significant
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40 pathological change. Gallium scintigraphy showed no hot lesions. Autoimmune
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43 pancreatitis was ruled out by magnetic resonance pancreatic cholangiopancreatography.
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46 Then, we performed pericardiocentesis by fine needle aspiration on 9 days after
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49 admission. The specimen was light yellow, cloudy, TP 5.2 g/dL, albumin 3.0 g/dL,
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52 IgG 1604.3 mg/dL, IgG4 451 mg/dL and abundant small sized lymphocytes, rich in
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55 IgG4-positive cells (Fig 2). There was no malignancy. Culture of the pericardial
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3 fluid revealed no bacterial infection. Therefore, we diagnosed this case as
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6 IgG4-related disease and treated with prednisolone 50 mg/day. Then his edema was
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9 rapidly improved. One month after starting steroid therapy, the CT revealed
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12 disappearance of pericardial effusion and regression of retroperitoneal mass (Fig 1C and
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16 D).

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19 To our knowledge, this is the first case report of diagnosed by fine needle aspiration
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22 cytology of large pericardial effusion. The pathological characteristic of this disease is
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25 rich in IgG4-positive plasma cells with typical fibrosis or sclerosis in the tissue [1].
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28 IgG4-related disease sometimes shows retroperitoneal fibrosis (RPF) and caused
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31 hydronephrosis as in our case. A biopsy for the retroperitoneal mass is necessary to
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34 provide a definitive diagnosis, but is considerably invasive. The major reasons of
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37 pericardial effusion are idiopathic pericarditis, infection included tuberculosis and
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40 malignancy [2, 3]. Secondary to malignancy are frequently observed in lung cancer,
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43 breast cancer, and malignant lymphoma. Malignant lymphoma is considered of
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46 differential diagnosis in both RPF and pericardial effusion. Fine needle aspiration of
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49 pericardial effusion is less invasive compared to retroperitoneal biopsy. Although
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52 Sugimoto et al. reported IgG4-related disease caused constrictive pericarditis diagnosed
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55 by pericardiectomy [4], pericardial effusion is not a major complication. We showed
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3 the cytology of pericardial effusion was rich in IgG4-positive cells similar to serum,
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6 suggesting that IgG4-related disease caused pericardial exudation of plasma cell.
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9 Thus, it is necessary to consider the possibility of IgG4-related disease in the differential
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12 diagnosis of pericardial effusion. A pericardiocentesis will be useful to diagnose as
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16 IgG4-related disease.
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25 The authors of this manuscript have certified that they comply with the Principles of
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28 Ethical Publishing in the International Journal of Cardiology [5].
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3 **Figure legends**
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6 Figure 1.
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9 CT scans on admission showing marked pericardial effusion (A) and retroperitoneal
10 mass around the aorta (B). One month after pericardiocentesis and corticosteroid
11 therapy, these findings were apparently ameliorated (C and D). The white arrow
12 indicates the retroperitoneal lesions.
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25 Figure 2.
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28 Cytological specimen obtained by fine needle aspiration from the pericardial effusion,
29 showing marked infiltration of matured plasma cells. A and B: Hematoxylin and Eosin
30 staining (x100 and x200). C and D: Immunohistochemical staining for IgG4 (x100
31 and x200). E and F: Immunohistochemical staining for the plasma cell marker, VS38
32 (x100 and x200).
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Figure 1

**Before
treatment**



**After
treatment**

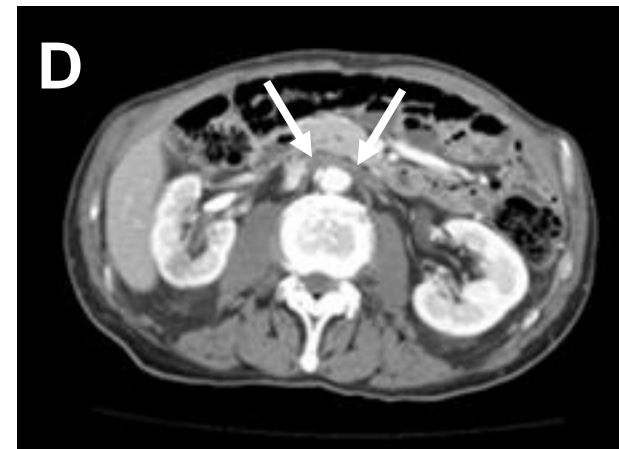


Figure 2

