Primary psoas abscess

Primary psoas abscess is a rare infection with an often vague and non-specific clinical presentation, especially in children. In Asia and Africa 99.5% of all psoas abscesses are primary, compared with 61% in the United States and Canada and 18.7% in Europe. Approximately 70% of psoas abscesses occur in patients younger than 20 years of age, with a male preponderance of 3:1. Fifty seven per cent of psoas abscesses occur on the right side, 40% on the left side, and 3% bilaterally. We present the following case and show the magnetic resonance imaging to emphasise the presenting signs, symptoms, and findings of this unusual infection.

A 13 year old white girl was in excellent health until she developed a dull ache in the inferior posterior thigh in association with fever to 38.9°C, nausea, vomiting, and diarrhoea. She walked with a limp. Her past medical history was non-contributory; she denied smoking, alcohol, drug use, or sexual activity. The girl was 1.52 m tall and weighed 70 kg. Vital signs were normal; temperature rose to 38.9°C within 24 hours of admission. A detailed general physical examination was normal. Abdominal and pelvic examinations were benign without organomegaly or peritoneal signs. Stool for occult blood was negative. Musculoskeletal examination was normal, with the exception of the left hip which showed pain on active and passive motion, particular abduction and medial rotation. The range of motion of the hip was normal; there was no localised warmth or palpation tenderness. The gait was antalgic for the left leg.

Laboratory examination showed a white blood cell count of 15.2 × 10^9/l (77% neutrophils/14% lymphocytes/8% monocytes) and platelets were 415 × 10^9/l. Erythrocyte sedimentation rate was 235 mm/h (normal <20 mm/1st h). Urine analysis disclosed trace blood and protein; the remainder of the laboratory tests were within normal limits. Blood and cervical cultures were negative. Posteroanterior radiographic examination of the left hip was normal. Bone scan was normal. Magnetic resonance imaging (MRI) of the abdomen and pelvis showed grossly abnormal signal intensities of the left psoas muscle (figs 1 and 2). Although a discrete abscess was identified, fine needle aspiration under imaging guidance, and microbial culture of the causative organism. If abdominal CT or MRI is unavailable, ultrasonography may demonstrate the inflammatory mass. Gallium-67 scanning may be useful in the diagnosis of psoas abscesses and detection of concomitant infectious foci.1–3 Differential diagnoses of psoas abscess include bacterial infection of the hip, avascular necrosis of the hip, irritable hip, necrotising fasciitis of the psoas muscle, pyelonephritis, pelvic inflammatory disease, retrocaecal appendicitis, pelvic disc, avascular necrosis, vertebral or pelvic osteomyelitis, and epidural abscess.1–3 These entities should be distinguishable upon the correlation of history, physical examination, laboratory tests, and imaging studies.

The cause of primary psoas abscess remains uncertain. Proposed mechanisms of psoas abscess formation include haematogenous spread from primary infectious foci or local trauma with intramuscular haematoma formation predisposing to abscess development.4 In secondary psoas abscess the most commonly associated disorder is Crohn’s disease; other disorders include ankylosing spondylitis, colonic inflammatory or neoplastic disease, and a variety of intra-abdominal or retroperitoneal infections.5–7 Primary psoas abscesses are caused by a single organism in 87.3% of cases: predominantly Staphylococcus aureus (88.4%), streptococci (4.9%), and Escherichia coli (2.8%). Blood cultures are positive in 41.7%, usually for Staphylococcus aureus.6 In the past decade the majority of patients with a primary psoas abscess were intravenous drug users (86%) infected with the human immunodeficiency virus (57%).7 Treatment for primary psoas abscess includes percutaneous drainage combined with systemic antibiotic administration.8 Surgical drainage is preferred for the patients in whom the psoas abscess is associated with underlying bowel disease.9 With appropriate treatment, psoas abscess rarely results in death (2.5%).10 Death from psoas abscess is associated more commonly with inadequate or delayed drainage, or both.10 Our patient responded well to antibiotic treatment and recovered completely.

Figure 1 Coronal magnetic resonance imaging scan of the abdomen showing abnormal signal intensity in the inferior pole of the left psoas muscle (arrows). Figure 2 Cross sectional magnetic resonance imaging of the pelvis showing abnormal signal intensity of the psoas muscle closely approximating the bladder (arrows).
Klippel-Feil syndrome in the prehispanic population of El Hierro (Canary Islands)

Klippel-Feil syndrome is an uncommon anomaly (1:40,000 births), mainly affecting the cervical spine.1 The classic clinical triad described by Klippel and Feil in 1912—short neck, low dorsal hairline, and restricted neck mobility—is the result of the fusion of a variable number of cervical vertebrae, sometimes reducing their number, and cervical spine bifida.2 Extraosseous changes,3 hemivertebra, vertebral body clefts, and thoracolumbar hemivertebrae,4 are sometimes seen.

In a prehispanic ossuary containing remains of at least 121 subjects in the island of El Hierro (Canary Islands), we noted:

1 A C2-3 block, with the two vertebrae fused both by the vertebral bodies and the medial ends of the arches, well preserved right intervertebral foramina, and foramina transversaria (fig 1), and a normal medullary canal. The body of the third vertebra shows intense degenerative signs.

2 A C5-6 block, consisting of two vertebrae with intense degenerative changes fused both by the vertebral bodies and the medial part of the arches. Both cervical blocks seem to belong to the same subject.

3 A well preserved atlas bone, with an incomplete anterior part of the left arch, with sharp, fine proximal, and distal ends (fig 2A), supporting an underlying developmental defect rather than an acquired one.

A left hemiatlas (fig 2B); although it is possible that the right body of the bone was partially fused with the left one (and was not recovered in the archaeological excavation), the posterior end of the arch was neither fractured nor fused to any other bony structure, thus pointing to a developmental defect.

Fusion of C2 and C3 (and C5-6), hypoplasia of the arch of the atlas,5 and complete bipartition of the atlas’ constitute distinctive features of Klippel-Feil disease. Thus the subject with the fused C2-3 and C5-6 blocks and the hemiatlas was probably affected by this disease. Possibly, the second atlas belongs to another subject with the same disease, though this possibility should be cautiously admitted. The two atlas bones show different developmental abnormalities. In the newborn, the ossification of the cartilaginous anterior and posterior arches of the atlas takes place progressively from the already ossified lateral masses. Often, especially in the anterior arch, secondary ossification centre(s) appear.6 In our case it seems that hypoplastic development of the anterior left arch took place. Because the hypoplastic part of the arch is in its middle part, probably, a second ossification centre was present, but ossification was never completed; in this sense, it is similar to the case described by Chigira et al,7 which also showed fusion of C5-6.

The “hemiatlas” perhaps is really an atlas with a midline cleft and a lost half, though the posterior arch does not reach the midline, so it never became fused with the right half of the bone. A secondary posterior ossification centre sometimes appears during the first years of life. In this case, it was absent, in contrast with the anterior secondary ossification centre which was surely present in the former case.

Perhaps familial links existed between the two subjects. Klippel-Feil syndrome is a heterogenous disorder, showing different alterations in different families.8 The simultaneous finding of different developmental abnormalities of the atlas in our two cases—assuming that the second one truly represents a case of Klippel-Feil—may either reflect an unusually high prevalence of this entity in the prehispanic population of El Hierro, or may also indicate that even in the same family clinical expression of the Klippel-Feil syndrome is variable.

of readjusting the immunological balance.\(^1\) As far as we know, only one case of primary Sjögren’s (SS) has been reported,\(^1\) with an unfavourable outcome. Another patient received an allogeneic bone marrow transplant and also had an unfavourable outcome.\(^1\) We describe here a further patient with primary SS who underwent HSCT for a non-Hodgkin’s lymphoma affecting the lung (large cell, mucosa associated lymphoid tissue (MALT) lymphoma) and review the literature on the effects of HSCT on the autoimmune features and histopathological changes in primary SS.

**Case report**

A white woman, aged 42, developed recurrent parotid swelling and symptoms of Sjögren’s (SS) has been reported,\(^1\) with an unfavourable outcome. Another patient re-

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### Table 1

<table>
<thead>
<tr>
<th>Variables</th>
<th>Patient 1</th>
<th>Patient 2</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age (years)</strong></td>
<td>57</td>
<td>Patient 2</td>
</tr>
<tr>
<td><strong>Diagnosis</strong></td>
<td>NHL* of the lung (large cells MALT* lymphoma)</td>
<td>Immunoblastic B lymphoma</td>
</tr>
<tr>
<td><strong>First line treatment</strong></td>
<td>6 cycles of F-MACHOP</td>
<td>VACOP-B* followed by VIPF=*</td>
</tr>
<tr>
<td><strong>Conditioning regimens</strong></td>
<td>ARA-C* 200 mg x 2/day x 4</td>
<td>BCNU 300 mg/m(^2)</td>
</tr>
<tr>
<td></td>
<td>VP16* 200 mg x 2/day x 4</td>
<td>Etoposide 800 mg/m(^2)</td>
</tr>
<tr>
<td></td>
<td>Cytarabine 1500 mg/m(^2)</td>
<td>Cytarabine 1600 mg/m(^2)</td>
</tr>
<tr>
<td></td>
<td>BCNU* 200 mg</td>
<td>Melphalan 140 mg/m(^2)</td>
</tr>
<tr>
<td><strong>Reinfusion</strong></td>
<td>MNC* 4.5 \times 10(^9)/kg</td>
<td>MNC 2.42 \times 10(^9)/kg</td>
</tr>
<tr>
<td></td>
<td>CD14 0.9 \times 10(^9)/kg</td>
<td>CD3 44.6 \times 10(^9)/kg</td>
</tr>
<tr>
<td></td>
<td>CD3 not counted</td>
<td>CD3 37.0 \times 10(^9)/kg</td>
</tr>
<tr>
<td>Literature on the complete continuous lymphoma remission after 3 years</td>
<td>Days to PMN* \times 10(^9)/l = 10 days</td>
<td>Days to PMN* \times 10(^9)/l = 11 days</td>
</tr>
<tr>
<td><strong>Sides effects/outcomes</strong></td>
<td>Alive in complete continuous lymphoma remission after 3 years</td>
<td>No remission of autoimmune disease</td>
</tr>
</tbody>
</table>

\(^*\)NHL = non-Hodgkin’s lymphoma; MALT = mucosa associated lymphoid tissue; F-MACHOP: F = 5-fluorouracil, M = methotrexate, A = Adriamycin, C = cyclophosphamide, H = doxorubicin, O = Oncovin, P = prednisone; ARA-C = arabinoside-C; VP16 = etoposide; CTX = cyclophosphamide; BCNU = carmustine; MNC = mononuclear cells; PMN = polymorphonuclear cells; Plt = platelets; VACOP-B: V = Vepesid, A = Adriamycin, C = cyclophosphamide, O = vincristine, P = prednisone, B = bleomycin; VIPF = vincristine, P = prednisone, F = etoposide.

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In table 2 we give the characteristics of the three major rheumatological autoimmune diseases treated with autologous stem cell transplantation.

### Table 2

<table>
<thead>
<tr>
<th>Autoimmune disease</th>
<th>Number</th>
<th>Died (No (%))</th>
<th>Transplant related death (No (%))</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systemic sclerosis</td>
<td>33</td>
<td>13 (39)</td>
<td>9 (27)</td>
</tr>
<tr>
<td>SLE</td>
<td>14</td>
<td>2 (14)</td>
<td>2 (14)</td>
</tr>
<tr>
<td>RA</td>
<td>35</td>
<td>1 (3)</td>
<td>1 (3)</td>
</tr>
<tr>
<td>Sjögren’s syndrome</td>
<td>2</td>
<td>1 (50)</td>
<td>0</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>84</td>
<td>17 (20)</td>
<td>12 (14)</td>
</tr>
</tbody>
</table>

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5 Euler HH, Marmont AM, Bacigalupo A, Fastenrath S, Dreger P, Hofmacht M, et al. G FERRACIOLI R DAMATO S DE VITA Division of Rheumatology, Udine Medical School, University of Udine, 33100 Udine, Italy pf.ferracioli@med.uniod.it gianfranco.ferracioli@dpmsc.uniud.it

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Castleman’s disease

A 65 year old woman presented in February 1998 with joint pains, mild weight loss, and a low grade irregular fever. Initially, pain was localised around both shoulder joints. Subsequently, elbows, wrists, hips, and knees were affected, with morning stiffness, but without particular morning stiffness. The symptoms were accentuated by movement, but also persisted during the night, often keeping the patient awake. On clinical examination, there was limited painful movement of the shoulders and hips with a marked reduction in strength. The small joints of the hands and feet were not affected. No other pathological conditions were found. Laboratory findings showed a marked increase in erythrocyte sedimentation rate (ESR, >100 mm/1st h), hyper-albuminemia and a mild anaemia, whereas enzymatic activity (serum aspartate aminotransferase, serum alanine aminotransferase, alkaline phosphatase, lactate dehydrogenase, and creatine kinase) was within the normal range. A diagnosis of polyglandular autoimmunity was made and a rapid and marked clinical improvement was observed with low dose steroid treatment (prednisone 12.5 mg/day). Pain disappeared, muscle strength and joint function became normal within a week. A decrease in ESR (to 40 mm/1st h) and α1 globulinaemia was noted after one month. The clinical condition remained satisfactory during 1998, with a complete normalisation of ESR and α1 globulinaemia after three months.

In January 1999 she developed jaundice and pruritus and a subcontinuous fever (38.5–39°C). Axial tomography and nuclear magnetic resonance disclosed compression of the biliary duct by a compact, well defined retroperitoneal mass (about 4 cm in diameter). Laboratory findings were typical of cholestasis (hyperbilirubinemia, increased serum alkaline phosphatase activity). Endoscopic retrograde colangiopancreatography showed slight enlargement of the biliary tract, dilatation of the gall bladder walls, and absence of gall stones. Transit of contrast medium along the biliary tract slowed down.

Explorative laparotomy confirmed the presence of a mass compressing the common bile duct. This was removed and histological examination disclosed a lymph node architecture characterised by evident follicular hyperplasia. Some of the germinal centres were enlarged and comprised polymorphous follicular centre cells, whereas other germinal centres were depleted of lymphocytes, and consisted predominantly of dendritic reticular cells showing vascular proliferation. The mantle zone was expanded and concentrically arranged around the atrophic germinal centres (onion skin layers). The interfollicular areas were also prominent, containing small lymphocytes, occasional cosinophils, plasma cells, and some immunoblasts, and showing numerous hyperplastic vessels of the post-capillary venous type. Occasionally these vessels, which were often hyalinised, penetrated the expanded follicles perpendicularly, giving rise to the so-called “lollipop” appearance (fig 1). An immunocytochemical study confirmed the normal organisation of the nodal structures, with a clear positivity of follicular elements for typical B cell markers CD20 and CD79a, and positivity of interfollicular lymphoid elements for T cell markers CD3 and CD45R0, whereas the dendritic reticular cells showed a typical positivity for CD21.

These features are distinctive of Castleman’s disease, also known by the descriptive term angiofollicular lymph node hyperplasia. This is a clinical entity characterised by angiofollicular hyperplasia of the lymph nodes without the presence of any atypical cells or other signs of malignancy. Many (multicentric or systemic form) or single (monocentric or solitary form) lymph nodal groups can be affected in the process and two histological subsets have been recognised: a hyaline-vascular type, characterised by marked expansion of the mantle follicle zone and a plasma cell type, with diffuse plasma cell proliferation in the interfollicular tissue. The cause of the disease is unknown, but overproduction of interleukin 6 has been shown in the course of Castleman’s disease and a possible pathogenic role for this cytokine has been suggested. It is commonly associated with several autoimmune conditions such as systemic lupus erythematosus, Behçet’s disease, amyloidosis, and with various neoplastic diseases, but its onset during the course of polymyalgia rheumatica has not been recently reported. No data exist about the possible relation between Castleman’s disease and polymyalgia, but a role for interleukin 6 can be suggested, perhaps through a stimulating action of this cytokine on monocytes and lymphocytes. The present case can be considered typical of the solitary form, hyaline-vascular type of Castleman’s disease. Our patient did not present any other lymphatic disease during the subsequent months, which would seem to confirm the good prognosis of this disease.

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