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 pole of the left psoas muscle (arrows). Note the proximity of the psoas wall with contrast medium injection. Definitive diagnosis is made by 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. Differentiation of psoas abscesses include bacterial infection of the hip, avascular necrosis of the hip, irritable hip, necrotising fasciitis of the psoas muscle, muscle, psoas abscess: worldwide variations in etiology. World J Surg 1986;10:834–43.

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Klippel-Feil syndrome in the prehispanic population of El Hierro (Canary Islands)

Klippel-Feil syndrome is an uncommon abnormality (1:40 000 births), mainly a heterogeneity disorder, showing different alterations in different families. The clinical expression of the Klippel-Feil syndrome is variable. 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 both the vertebral bodies and the medial ends of the arches, well preserved right intervertebral foramina, and foramina transversaria (fig 1), and a normal medullar 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 hemi-vertebra (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, 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 hemi-atlas 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. 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., which also showed fusion of C5-6.

The “hemi-vertebra” 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. 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

Figure 1  Fused C2-3 vertebral block.

Figure 2  (A) Atlas with a hypoplastic left arch; the lack of fusion of the anterior arch of the left foramen transversarium is also evident. (B) Left hemi-vertebra.
of readjusting the immunological balance. As far as we know, only one case of primary Sjögren’s (SS) has been reported, with an unfavourable outcome. Another patient received an allogeneic bone marrow transplant and also had an unfavourable outcome. 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 syndrome sicca, with a Schirmer’s test of 5 mm in the right eye and 4 mm in the left eye. Break up time was 6 s and sialometry was < 1 ml. She had periodic relapses of her parotid swelling. In August 1994 (aged 57) lung x-rays and computed tomography disclosed a parenchymal nodule of 3 cm in diameter in the basal left lobe. She underwent a lobectomy that disclosed a MALT of the large cell B lymphoma histotype, stage IE. In December 1994 two more nodules in the right lobe, with hilar bilateral adenomegaly, led to the diagnosis of a relapse of her lymphoma, which had progressed to stage IV. She then received six courses of F-MACHOP (vincristine 0.5 mg/m² at hours 0 and 12; cyclophosphamide 800 mg/m² intravenous (IV) bolus at hour 36, 5-fluorouracil 15 mg/kg IV for six hours at hour 36, cytosine-arabinoside 1000 mg/m² IV for six hours at hour 42, doxorubicin 60 mg/m² IV bolus at hour 48, methotrexate 500 mg/m² IV for six hours at hour 60, predonime 60 mg/m² from day 1 to 14), and follicin rescue (20 mg/m² IV bolus at hours 84, 96, 108, 120), with a prompt reduction of hilar adenopathies and a net decrease of pulmonary nodule size. However, no complete remission was recorded. She was then offered the possible chance of an HSCT. Table 1 reports the possible conditioning regimens or myeloablation that disclosed a MALT of the large cell B lymphoma histotype, stage IE. In December 1994 two more nodules in the right lobe, with hilar bilateral adenomegaly, led to the diagnosis of a relapse of her lymphoma, which had progressed to stage IV. She then received six courses of F-MACHOP (vincristine 0.5 mg/m² at hours 0 and 12; cyclophosphamide 800 mg/m² intravenous (IV) bolus at hour 36, 5-fluorouracil 15 mg/kg IV for six hours at hour 36, cytosine-arabinoside 1000 mg/m² IV for six hours at hour 42, doxorubicin 60 mg/m² IV bolus at hour 48, methotrexate 500 mg/m² IV for six hours at hour 60, predonime 60 mg/m² from day 1 to 14), and follicin rescue (20 mg/m² IV bolus at hours 84, 96, 108, 120), with a prompt reduction of hilar adenopathies and a net decrease of pulmonary nodule size. However, no complete remission was recorded. She was then offered the possible chance of an HSCT. Table 1 reports the possible conditioning regimens or myeloablation.

Table 1 Lymphoma characteristics, conditioning regimens, side effects and outcome of the two female patients so far studied, after haematopoietic stem cell transplantation

<table>
<thead>
<tr>
<th>Variables</th>
<th>Patient 1</th>
<th>Patient 2</th>
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<tbody>
<tr>
<td>Age (years)</td>
<td>Diagnosis</td>
<td>Conditioning regimen</td>
</tr>
<tr>
<td>57</td>
<td>NHL* of the lung (large cells MALT* lymphoma)</td>
<td>F-MACHOP</td>
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<tr>
<td>6 cycles of F-MACHOP</td>
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<tr>
<td>34</td>
<td>Immunoablative B lymphoma</td>
<td>VACOP-B* followed by VIFPE*</td>
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<tr>
<td></td>
<td></td>
<td>BCNU 300 mg/m²</td>
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<td></td>
<td></td>
<td>Etoposide 800 mg/m²</td>
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<td></td>
<td></td>
<td>Cycarabine 1600 mg/m²</td>
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<td></td>
<td></td>
<td>Melphalan 140 mg/m²</td>
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<td></td>
<td></td>
<td>MNC 2.42 ± 10⁹/kg</td>
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<td></td>
<td></td>
<td>CD34 4.60 ± 10⁹/kg</td>
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<td></td>
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<td>CD33 37.03 ± 10⁹/kg</td>
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<td></td>
<td></td>
<td>Days to PMN* &gt;10¹⁰/l = 11 days</td>
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<td></td>
<td></td>
<td>Days to Pi* &gt;20 g/dl = 8 days</td>
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<td>Literature on the continuous lymphoma remission of 3 years</td>
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<td>No remission of autoimmune disease</td>
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<td>Died 20 months after transplantation for Pneumocystis carinii pneumonia</td>
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<tr>
<td></td>
<td></td>
<td>Reinfusion</td>
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<td></td>
<td></td>
<td>CD14 0.9 ± 10⁹/kg</td>
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<tr>
<td></td>
<td></td>
<td>BCNU* 200 mg</td>
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<td></td>
<td></td>
<td>MNC* 4.5 ± 10⁹/kg</td>
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<tr>
<td></td>
<td></td>
<td>CD3 not counted</td>
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<td></td>
<td></td>
<td>Days to PMN* &gt;10¹⁰/l = 10 days</td>
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<td>Days to Pi* &gt;20 g/dl = 8 days</td>
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<td>Literature on the continuous lymphoma remission of 3 years</td>
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<td>Died 20 months after transplantation for Pneumocystis carinii pneumonia</td>
</tr>
</tbody>
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* 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 = predonime; ARA-C = arabinoside-C; VP16 = etoposide; CTX = cyclophosphamide; BCNU = carmustine; MNC = mononuclear cells; PMN = polymorphonuclear cells; Pi = platelets; VACOP-B: V = Vepesid, A = Adriamycin, C = cyclophosphamide, O = vincristine, P = predonime, B = bleomycin; VIFPE: V = vinblastine, I = ifosamide, P = predonime, E = etoposide.

In table 1 we give the characteristics of the other patient with primary SS (No 2), previously reported. It can be seen that the conditioning regimen used was identical, with the exception of the stem cell rescue, and bone marrow reconstitution were different. However, in this case, also, SS was not cured and there was no remission of the autoimmune reaction, which showed persistence of the immunological imbalance and poor function of the salivary organs.

Current data suggest that best results have been obtained in RA, the worst in SS, suggesting that the helper 2 oriented diseases have a poorer response.

Results for SS seem to confirm this because HSCT cured lymphoma but did not improve the autoimmune disease. No changes were recorded in the function of salivary glands, or in the synthesis of ANA, or the histopathology. The other case reported did show some early improvement. An immunological reassessment might thus have revealed a different outcome. It seems unlikely that T cell depletion would offer a better prospect, especially in view of the increased risk of long term immunosuppression, lymphoproliferative diseases, and infections.

On the other hand, allogeneic bone marrow transplantation, even though clearly appealing given the chance of eradicating the intrinsic stem cell defect, does not represent a definite cure either and the related morbidity-mortality still remains too high to be accepted as a possible alternative. As benefits have been seen in around two thirds of the cases treated so far, controlled trials in the three major rheumatic diseases are eagerly awaited.

We gratefully acknowledge the invaluable help we received from Professor A. Tyndall, who provided us with the latest data available of the EBMTC/EULAR registry on haematopoietic stem cell transplantation in autoimmune diseases.

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 partitional swelling. 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- 
| globulinaemia 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 polymyalgia rheumatica was made and a rapid and marked clinical improvement was obtained 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 \( \alpha_2 \) globulinaemia was noted and maintained. The clinical condition remained satisfactory during 1998, with a complete normalisation of ESR and \( \alpha_2 \) globulinaemia after three months.

In January 1999 she developed jaundice with 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 retropancreatic mass (about 4 cm in diameter). The laboratory pattern was typical 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 lympho-hyaline type 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, lympho-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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Haematopoietic stem cell transplantation (HSCT) in a patient with Sjögren's syndrome and lung malt lymphoma cured lymphoma not the autoimmune disease

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