limb ischaemia in a vascular laboratory if the clinical diagnosis of claudication is in doubt.

References

SHORT REPORTS

Primary hyperparathyroidism presenting with visual failure and palatal swelling

We report the case of a patient with primary hyperparathyroidism in which the presenting symptoms included unilateral visual failure. This recovered completely after removal of a parathyroid adenoma.

Case report
A 47-year-old housewife presented with a midline palatal swelling for 10 weeks. She also complained that vision in her right eye had been diminishing over four weeks. She had had sciatica two years previously and still had diffuse back pain. She was edentulous, wearing full upper and lower dentures. There was a smooth, fairly soft swelling 3 cm x 1.5 cm in the midline at the posterior aspect of the hard palate, displacing the upper denture, and two small fusiform swellings on the lateral aspects of the maxillary alveolus in the incisor regions. Slight localised bossing of the left front bone was apparent. Visual acuity in the right eye was very poor and deteriorated to perception of light over 10 days. Corneal calcification was noted bilaterally at the limbal equator. The retinal arteries in the right eye appeared slightly attenuated and the pupil was sluggish, but there was no lens opacity, myopia, proptosis, impairment of ocular movement, or papilloedema and no ocular lesion sufficient to account for the loss of vision. Visual acuity in the left eye was normal. X-ray examination showed ill-defined radiolucent areas in the maxillary alveolus coinciding with the fusiform swellings. The right optic foramen appeared on tomograms as an ill-defined region of radiolucency with no clear margin. The left optic foramen appeared normal. The phalanges showed subperiosteal erosions. There were cystic changes in the pelvic girdle, ribs, and frontal bone.

Serum concentrations measured were as follows: calcium 3.85 mmol/l (15-4 mg/100 ml) (normal 2.1-2.6 mmol/l [8-4-10.4 mg/100 ml]); phosphate 0.72 mmol/l (2.3-5 mg/100 ml) (normal 0.8-1.4 mmol/l [2.5-4.5 mg/100 ml]); albumin 45 g/l (normal 34-50 g/l); alkaline phosphate 303 IU/l (normal 20-90 IU/l); parathyroid hormone 6.65 μg/l (normal <1.0 μg/l) by radioimmunoassay using a human parathyroid hormone standard; creatinine 91 μmol/l (1.03 mg/100 ml) (normal 50-120 μmol/l [0.56-1.35 mg/100 ml]).

An excision biopsy specimen of the palatal swelling showed a vascular, fleshy tumour completely eroding the bone of the hard palate, macroscopically typical of a giant-cell lesion. Histologically it was a giant-cell lesion containing a large amount of osteoid, some showing early ossification. A large single tumour of the left inferior parathyroid gland weighing six grams was removed. Histologically it was typical of a parathyroid chief-cell adenoma. Postoperatively all the biochemical abnormalities resolved. She was given 0.5-2 μg 1-alpha-hydroxycholecalciferol daily for 12 weeks, by which time the alkaline phosphate concentration had returned to normal. Radiological healing of the skeletal lesions was subsequently confirmed. In particular, the right optic foramen became sharply defined, normal in size, and with a slightly sclerotic border. Vision returned in the right eye within two weeks of the parathyroidectomy. Acuity was 6/9 after six weeks and 6/5 after five months.

Comment
This patient had skeletal changes affecting many bones because of primary hyperparathyroidism. The palatal giant-cell tumour was typical of lesions described in this condition and well known to affect the jaw bones. 1 Corneal calcification is associated with hypercalcaemia and papilloedema has been reported, but visual failure due to hyperparathyroidism has not been recorded. It may have resulted from distortion of the neurovascular bundle in the optic canal by another giant-cell lesion, and this hypothesis is supported by the return of vision and restoration of the optic foramen (radiologically) that followed removal of the parathyroid adenoma.

We thank Drs A R Manners, E Dillon, and A K Lamballe for radiological advice, Drs R Lawler and H E Simpson for histological reports, and the Suprasurgical Assay Service (Middlesex Hospital Laboratory, London) for measurements of parathyroid hormone.


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Chronic thrombocytopenic purpura associated with toxoplasmosis

Many aetiological factors play a part in the pathogenesis of idiopathic thrombocytopenic purpura. Among these are viral infections, drugs, and other autoimmune diseases. Many cases are truly idiopathic. We report the case of a child with chronic thrombocytopenic purpura associated with toxoplasmosis. The parasite Toxoplasma gondii has not been reported as a possible cause of thrombocytopenic purpura.

Case report
A 2-year-old girl presented with purpura. One month earlier she had had a fever, but the cause was not known. Physical examination showed purpura, enlarged lymph nodes, and mild hepatosplenomegaly. The platelet count was 10-20 x 10^9/l (10 000-20 000/mm^3). The result of bone marrow examination was unhelpful. After a short course of steroid treatment the purpura and thrombocytopenia disappeared. A Sabin-Feldman dye test, performed because of an enlarged spleen and liver and lymphadenopathy, was positive at a titre of 1/4096. Three months later the titre was 1/1024, and one year later 1/256.
Three months after the first episode of purpura she presented again with diffuse purpura. The platelet count was $6 \times 10^{10}$ (6000/mm$^3$), and the results of examination of the bone marrow was consistent with the diagnosis of idiopathic thrombocytopenic purpura. She had clearly had a relapse. The results of tests for LE cells, antinuclear factor, and red-cell antibodies were negative. Cellular and humoral immunity function tests were normal. The thrombocytopenia and purpura persisted despite treatment with steroids and azathioprine, so a splenectomy was performed when she was 6 years old. After this she had a complete clinical and haematological recovery.

Comment

All but 5% of patients with thrombocytopenic purpura are considered to have the idiopathic form, in which no preceding infection can be shown. One-half of this group have the true idiopathic form and the other half the postviral idiopathic form. Possibly, therefore, a patient who is thought to have the true idiopathic form might move to the postviral group when an infectious illness can be shown to have preceded the onset of purpura. This may have been the case with our patient. She fulfills the criteria proposed by Feldman for the diagnosis of toxoplasmosis. A titre of 1/4096 in the dye test and a rising titre in two serum specimens are compatible with the diagnosis of a recently acquired infection.

Toxoplasma infection is much more common in people with compromised immunological states. This raises the possibility that both the idiopathic thrombocytopenic purpura and the toxoplasmosis in our patient resulted from an altered immune mechanism that was not detected by standard laboratory procedures. Cell-mediated immunity may play a part in the pathogenesis of idiopathic thrombocytopenic purpura. Recently, blast transformation of lymphocytes in the presence of autologous platelets was shown. Is the production of antiplatelet antibodies mediated through the inhibition of suppressor T cells? Or are there two parallel mechanisms: one of cell-mediated immunity and the other of humoral immunity? Is toxoplasma infection the result of an altered immune state, or did the parasite cause such a change in cell-mediated immunity that idiopathic thrombocytopenic purpura appeared? These and other questions await an answer.

Interestingly, we could not detect autoantibodies in the sera of our patient after splenectomy. In our laboratory we found a significant increase in the incidence of autoantibodies in patients who have had splenectomies. In view of our patient’s condition, we suggest that the term “postviral idiopathic thrombocytopenic purpura” should be extended to “postinfectious idiopathic thrombocytopenic purpura” to include the possible infectious agents that, although they are not viruses, may play a part in causing thrombocytopenic purpura.

Primary thrombocytopenia in monzygotic twins

The aetiology of myeloproliferative disorders is unknown but there is some evidence that genetic factors may play a part. Although there are several reports of familial polycythaemia there are very few of primary thrombocytoma. This report is of primary thrombocythaemia in monzygotic twins.

Case report

A 35-year-old man (A) had lost 6 kg in weight when he presented with a 10-week history of partial blinding in one eye. He had a right subhyaloid haemorrhage and his spleen was enlarged 7 cm below the costal margin. He was anaemic, and had a platelet count of 954 $\times 10^9$ (954,000/mm$^3$). His asymptomatic twin brother (B) had a radiologically enlarged spleen and a platelet count of 1330 $\times 10^9$ (1,330,000/mm$^3$) but he was not anaemic. The full blood counts are given in the table. B’s blood film was morphologically normal but that of A showed polikyocytes, teardrop cells, and target cells. Their bone marrows yielded only cells of the normal male karyotype.

The morphology of marrow smears and bone trephines from both men were similar. The smears contained no stainable iron, erythropoiesis was normoblastic, and there were dyserythropoietic changes. There were sheets of platelets and numerous megakaryocytes. There was no increase of reticulin and no fibrosis in the bone trephines. All other investigations, including measurement of the leucocyte alkaline phosphatase concentration, were normal except that A had hyperuricaemia, with uric acid concentration 0.59 mmol/l (10 mg/100 ml) (normal 0.15-0.42 mmol/l (2.5-7.0 mg/100 ml)).

Blood counts in monzygotic twins with primary thrombocythaemia

<table>
<thead>
<tr>
<th>Age when tested (years)</th>
<th>Haemoglobin (g/dl)</th>
<th>Mean cell vol (fl)</th>
<th>White cells (10$^9$/l)</th>
<th>Platelets (10$^9$/l)</th>
<th>Red cells (10$^9$/l)</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>35</td>
<td>11.8</td>
<td>87</td>
<td>12.4</td>
<td>954</td>
</tr>
<tr>
<td>B</td>
<td>35</td>
<td>11.7</td>
<td>86</td>
<td>12.3</td>
<td>954</td>
</tr>
<tr>
<td>Father</td>
<td>67</td>
<td>11.4</td>
<td>93</td>
<td>6.7</td>
<td>328</td>
</tr>
<tr>
<td>Mother</td>
<td>65</td>
<td>11.7</td>
<td>90</td>
<td>8.5</td>
<td>250</td>
</tr>
<tr>
<td>Sister</td>
<td>37</td>
<td>12.3</td>
<td>88</td>
<td>6.3</td>
<td>235</td>
</tr>
<tr>
<td>Daughter (of A)</td>
<td>15</td>
<td>12.7</td>
<td>90</td>
<td>7.8</td>
<td>390</td>
</tr>
<tr>
<td>Son of A</td>
<td>12.3</td>
<td>84</td>
<td>90</td>
<td>8.6</td>
<td>390</td>
</tr>
<tr>
<td>Son of B</td>
<td>7</td>
<td>11.8</td>
<td>81</td>
<td>7.4</td>
<td>350</td>
</tr>
</tbody>
</table>

Conversion: SI to traditional units—Mean cell volume: 1 fl = 1 μm$^2$.

The red cell genotype of the twins and their close relations together with tissue typing studies suggested that the twins were monzygotic. None of their relations had thrombocythaemia. The twins were both treated for primary thrombocythaemia. After initial treatment with radioactive phosphorus the platelet count of both men fell below 400 $\times 10^9$/l (400,000/mm$^3$) within four months. By then A had gained 7 kg and his haemoglobin concentration had risen to 13.6 g/dl. During the subsequent six years neither has developed polycythaemia but both have had several courses of busulphan when their respective platelet counts rose above 400 $\times 10^9$/l.

Comment

Dedichen et al. in a paper describing nine patients with haemorrhagic thrombocythaemia included a 40-year-old woman with five similarly affected siblings. The platelet count of four of the siblings was not available but the fifth, a man, had splenomegaly associated with thrombocythaemia. Beretta Anguissola and Prato described thrombocythaemia associated with gastrointestinal haemorrhage in three male members of three generations of an Italian family. Flickers and Speck described a 47-year-old man with primary thrombocythaemia. He had been taking thiopeta for two years when cytogenetic studies showed hypoploidy in 8 of 70 karyotypes. His sister, who had hepatosplenomegaly and thrombocythaemia, refused to have a marrow aspirate, but probably her brother’s chromosomal abnormalities were the result of cytotoxic therapy.

The present report is unique in that it describes primary thrombocythaemia in monzygotic twins. There is no evidence that any of their close relations has a similar disorder, but the presentation of the same myeloproliferative disease in the twins is evidence that a genetic factor may play some part in its pathogenesis.

I thank Professor S Pearl for referring the patient, Dr S Lawler for the cytogenetic studies, and Dr R Sanger for red cell genotyping.