References


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Wernicke aphasia and cardiac embolism

SIR: It has been suggested that Wernicke aphasia is usually due to cerebral (cardiac) embolism, with the implication that cardiac emboli cause infarction in different sites than are associated with carotid artery disease. To test this hypothesis we have looked at the clinical and CT evidence of the localisation of the sites of infarction in the dominant hemisphere of a group of dysphasic patients separated into a subgroup with presumed cardiac emboli and another with presumed carotid artery disease.

Dysphasic stroke patients were reviewed by a retrospective case note study. Cardiac embolism was assumed to be likely in those with atrial fibrillation (n = 27). Additional evidence of cardiac disease was present in seven of the patients (rheumatic heart disease in six, alcoholic cardiomyopathy in one). In a second subgroup of 67 cases a clinical diagnosis of ischaemic stroke causing dysphasia had been made but there was no evidence of cardiac disease and the patients were in sinus rhythm. More direct evidence of carotid artery disease was present in 33 of these patients from their angiography or Doppler studies.

The clinical and neuropsychological assessments were reviewed for evidence of the localising implications of the type of dysphasia detected. Broca, Wernicke, and global aphasia was recognised, as was a syndrome implying parietal lobe localisation (conduction aphasia with or without dysgraphia and dyscalculia or other elements of Gerstmann's syndrome). Patients with mild dysphasia could often not be classified and are listed as "unclassified".

CT scan reports were studied for evidence of the localisation of cerebral infarction in the dominant hemisphere.

The type of dysphasia encountered in the "cardiac" and "carotid" groups is set out in table I. Wernicke aphasia was noted in 15% (4/27) of the "cardiac" embolic group but in only 4-5% (3/67) of the carotid disease cases. The difference is not, however, significant at conventional levels (Chi squared 3-2 p < 0-07).

CT scans were available in 16 of the "cardiac" group and 38 of the noncardiac or carotid group. The siting of the infarcts in the two groups is shown in table 2. A temporal lobe lesion was found in 31% (5/16) of the scanned cardiac cases, but in only 3-5% (2/58) of the carotid group (Chi squared with Yates correction 11-6 p < 0-01).

This review of the clinical picture of the type of dysphasia encountered in stroke patients has suggested that there is a difference between the effects of cardiac embolism and the infarction due to presumed carotid artery disease. Wernicke aphasia was more common in the group of patients with atrial fibrillation whose cerebral infarct was assumed to be due to cardiac embolism. The CT scan data also revealed temporal lobe infarction to be more common in the "cardiac" group.

This small study thus tends to confirm the clinical suspicion that embolic infarction in the temporal lobe with appropriate clinical sequelae is more frequent when emboli

<table>
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<th>Type of dysphasia in infarcts of different aetiology</th>
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<tr>
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<td>Global</td>
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<tr>
<td>1. Cardiac (A.Fib.)</td>
<td>11</td>
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*Chi squared 3-2.

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<th>CT scan localisation of infarcts of different aetiology</th>
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<td>Normal</td>
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<tr>
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<tr>
<td>2. Carotid</td>
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*Chi squared 11-6 p < 0-01.
Chronic relapsing inflammatory polyneuropathy complicating sicca syndrome

Sirs: Sjögren’s syndrome is a multi-system autoimmune disease. The cardinal features of dry eyes and mouth are caused by a destructive mononuclear cell infiltration of the lachrymal and salivary glands. By themselves, these features are thought of as the sicca syndrome whereas involvement of other organs systems, in a connective tissue disease, comprises Sjögren’s syndrome. Lesions of the peripheral and central nervous systems are described in both syndromes but are rare. In this report a case of the sicca syndrome is described associated with chronic relapsing inflammatory polyneuropathy.

A 64 year old caucasian woman was diagnosed in 1977 as having sicca syndrome. She had the characteristic dry eyes and dry mouth. A Schirmer tear test showed a marked reduction of tears in both eyes. She was also found to have pernicious anaemia with the demonstration of gastric parietal cell antibodies positive at 1:10 and positive intrinsic factor antibodies. Thyroid, thyroglobulin and microsomal antibodies were also positive. Antinuclear antibodies were negative. In November 1981 she developed generalised weakness in her legs. There was a mild improvement initially, followed by a marked deterioration during February 1982. This relapse improved spontaneously a few weeks after its onset. She continued to improve until October 1982. She then suffered an acute deterioration in leg power. During November 1982 her arms became involved and she was unable to brush her hair. Over the next 2 to 3 months she became unable to walk. Examination at this time showed mild bilateral facial weakness, weakness of the trapezius muscles bilaterally with weakness of neck flexion. She had a generalised reduction in power, especially proximally with asymmetric sluggish reflexes and flexor plantar responses. The CSF protein was elevated at 0·8 g/l without cells and there was no oligoclonal pattern. Nerve conduction studies showed a median distal motor latency of 6·5 ms with a motor conduction velocity from the elbow to the wrist of 24 m/s. Using a surface electrode over the right extensor digitorum brevis muscle, there was a distal motor latency of 7·0 ms and a common peroneal nerve motor conduction velocity from knee to ankle of 29 m/s. A sural nerve biopsy showed a moderate reduction in the myelinated nerve fibre population. There was no evidence of active nerve fibre breakdown, demyelination or remyelination but there were moderately numerous regenerative clusters. A muscle biopsy performed at the same time was consistent with chronic partial denervation.

She was treated with prednisolone 1 mg/kg and azathioprine 2 mg/kg daily. Over the next 3 months she improved, but even on this therapy, she then relapsed becoming rapidly weaker. During the first week of March 1983, she became tetraplegic and suffered respiratory embarrassment with a vital capacity between 800 and 1300 ml. At this time, whilst deteriorating, she received a course of five plasma exchanges. Each exchange removed three litres of plasma. After the first exchange she was stronger and in particular, there was improved movement in the hands and fingers. With each subsequent exchange there was further recovery in ventilatory function and limb power. She was continued on prednisolone 1 mg/kg and azathioprine 2·5 mg/kg on which she was maintained over the next 18 months, when with full recovery the therapy was reduced and subsequently stopped. Up to now there has been no further relapse.

The patient reported had a relapsing polyneuropathy demonstrated by the reduced motor conduction velocities to be demyelinating in type. Peripheral neuropathies have been described in Sjögren’s and the sicca syndrome but these cases were mainly sensory distal axonopathies. Vincent et al reported the case of a 53 year old woman with six episodes of recurrent cranial nerve palsies, the diagnosis of Sjögren’s syndrome being made 7 years after the first episode. Others have reported on cranial neuropathies, in particular sensory trigeminal neuropathy. Mixed and asymmetric neuropathies together with monoclonal neuritis multiplex can also occur but these are uncommon. Marbini et al have reported on a case with chronic progressive hypotrophic neuropathy. They comment on the histological similarities between their case and some reported cases with chronic relapsing polyneuropathy. Both chronic relapsing polyneuropathy and Sjögren’s syndrome have an HLA association and it is surprising that no previous cases have been reported. Cases of chronic relapsing inflammatory neuritis are associated with HLA A1, B8, DRW3 haplotype and in Sjögren’s syndrome there is an association with the HLA B8, DW2 and DW3 haplotype or associated with rheumatoid arthritis, HLA D4. A recent report included five out of eight patients with chronic demyelinating inflammatory polyneuropathy who had associated autoimmune disease. This lends support to the notion that the disease may have an autoimmune basis though other series have not had such a clear association.

The mononeuritis may be the result of vasculitic damage to the vasa nervorum though direct infiltration of nerve sheaths with mononuclear lymphocytes may also produce a neuropathy. The sural nerve and muscle biopsies in this case failed to reveal either vasculitis or lymphocytic infiltration. The patient responded well to the combined therapy of immunosuppression and plasma exchange. This was in keeping with a recent study, including this case, where immunosuppressive therapy with prednisolone (1 mg/kg), azathioprine (2·0–2·5 mg/kg) and plasma exchange was found to be more effective than immunosuppressive therapy alone. A retrospective analysis of cases has shown that corticosteroid treatment was of little benefit in the CNS complications of Sjögren’s syndrome. Steroids may however be of benefit where the peripheral nervous system involvement is more marked and clearly vasculitic, but this was not the case in two out of three patients recently reported. Previous publications have commented on the similarity of Sjögren’s syndrome to SLE in the involvement of the nervous system. This patient could not meet the minimum clinical criteria for having SLE, though the presence of chronic relapsing inflammatory polyneuropathy would also be uncommon in this disease.

I am grateful to Professor PK Thomas for permission to report this case.

References

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M J Harrison and J Marshall

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