Paget’s Disease in Diabetic Subjects

Sir,

Paget’s disease or osteitis deformans is a chronic disorder of unknown etiology, resulting in replacement of compact bone by vascular spongy bone due to excessive breakdown and formation of disorganized bone tissue. The probable causes are abnormal gene expression or viral infection. Paget’s disease is diagnosed on the basis of radiological evidence (X-rays and bone scan) and biochemical parameters (elevated serum alkaline phosphatase levels). Paget’s disease is more prevalent in Western countries where the prevalence is around 3 in people over 40 years of age and the male to female ratio is 3:2. It is considered to be rare in Asian countries.

An earlier study on Paget’s disease revealed that the prevalence of diabetes among affected individuals was lower when compared to those unaffected with Paget’s disease. However, both diabetes mellitus and Paget’s disease are metabolic disorders. Evidences suggest that bone metabolism is altered in diabetic subjects. Although, there is no literary documentation of any association between these two disorders, both the diseases have been hypothesized to have an immunological and genetic basis. In our clinical practice, we observed that many diabetic patients visiting our centre, had unexplained elevated levels of serum alkaline phosphatase (ALP). Hence, we conducted a retrospective study to assess the prevalence of Paget’s disease among diabetic subjects registered at our centre in order to evaluate any presence of any association between the two diseases. The study protocol is outlined in Fig. 1.

Out of 28732 (18930 males and 9802 females) consecutive diabetic patients who had liver function tests (LFT) done at our centre, 298 (198 males and 100 females) had ALP levels ≥500 IU/L (the normal value for ALP in our lab is 350 IU/L). Of these 298 patients, 87 (60 males and 27 females) had isolated elevated serum ALP levels with no other abnormal biochemical changes in LFT. On reviewing the case details of these 87 subjects, 19 (15 males and 4 females) were found to be known cases of Paget’s disease undergoing treatment for the same. These patients were diagnosed by an endocrinologist to have Paget’s disease on basis of persistently elevated serum ALP levels, X-rays and bone scans. These confirmed cases have been treated with Inj. Palmidronate and are being followed up by the endocrinologist and they are doing well. The prevalence of known Paget’s disease in the total diabetic population visiting our center is therefore, 19/28732 (0.066%), which includes 79% males (n=15) and 21% females (n=4). While categorizing according to age, it was interesting to note that all the subjects with known Paget’s disease were above 50 years of age.

Though the remaining 68 patients (45 males and 23 females) with elevated ALP levels had no further investigations to confirm Paget’s, one cannot rule out the possibility that some of them may be affected by Paget’s disease. Other causes of isolated elevated serum alkaline phosphatase levels include chronic active hepatitis, obstructive jaundice, pregnancy, bone disease, drug toxicity and inflammatory bowel disease. However, none of these patients had any clinical evidence of these conditions.

We would like to suggest that the Paget’s disease of the bone should be looked for in elderly diabetic patients, especially in those who have isolated elevation of serum ALP levels.

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It has presented under various guises of entirely unrelated disorders such as tuberculosis, sarcoidosis, sporotrichosis, molluscum contagiosum, tuberous xanthomas etc.\textsuperscript{1,2} Similarly, nerve abscesses that result from focal necrosis of the leprous granuloma have also been misdiagnosed, especially under unfamiliar settings, by general physicians, surgeons or neurologists, leading to serious and irreparable consequences such as muscle palsies and limb deformities. We report an interesting episode of greater auricular nerve abscess with enlargement of transverse (anterior) cutaneous nerve, misdiagnosed as external jugular vein thrombosis in a surgical setting.

A 30 years male presented to Cardiothoracic and Vascular Surgery department with a cord-like swelling on the left side of neck since 5 months and pain in the swelling for last one week. There were no complaints of infective process in the head and neck region, trauma or surgical intervention. A diagnosis of external jugular vein thrombosis substantiated by venous doppler of left neck vessels, was made. The coagulation profile was normal. Surgical intervention was planned and the patient referred to dermatology department for evaluation of an erythematous plaque on the corresponding cheek.

Dermatological examination revealed an irregular, 3 x 4 cm sized, erythematous, well-defined hypoesthetic plaque on the left cheek. The left lateral aspect of neck showed two, firm, cord-like swellings that formed a V-shaped configuration with one linear cord running from midway of the sternomastoid muscle to the submandibular region and the other perpendicular to it, the apex being formed by a globular, 2x1 cm, erythematous, fluctuant, tender swelling (Fig. 1). FNAC of the globular swelling showed acute and chronic inflammatory infiltrate with multiple Langhan giant cells and necrosis, within which few acid-fast bacilli (AFB) could be identified. Skin biopsy from the cheek plaque was consistent with borderline tuberculoid leprosy. Nerve biopsy of the greater auricular nerve showed numerous epithelioid cell granulomas with few

<table>
<thead>
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<th>Table 1: Differences between greater auricular nerve abscess and external jugular vein thrombosis</th>
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<td><strong>Anatomy</strong></td>
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<td>Arises from posterior triangle, of the lateral border of sternomastoid muscle, surrounds and then traverses it obliquely towards the ear</td>
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<td><strong>Clinical manifestations</strong></td>
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<td>Nerve abscess manifests as tender fluctuant nodule along the course of nerve with accompanying sensory deficit.</td>
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<td><strong>Diagnosis</strong></td>
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<td>Finding of hypoesthetic skin lesion with characteristic histopathology of skin and nerve. Demonstration of AFB.</td>
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fragmented AFB. A repeat venous doppler and MRI of the brain showed no evidence of venous thrombosis. A diagnosis of borderline tuberculoid leprosy in type I reaction, with greater auricular and transverse (anterior) cutaneous nerve enlargement and greater auricular nerve abscess was made.

The patient was started on paucibacillary antileprosy therapy with tapering doses of daily steroids, with significant reduction in erythema over the cheek lesion and pain and erythema over the abscess after two weeks. After three months, the plaque was barely visible, the swellings became less prominent and the abscess ruptured with intermittent pus discharge, for which incision and drainage was done.

Nerve involvement is a unique feature of leprosy due to the affinity of Mycobacterium leprae for schwann cells. Nerve abscess occurs commonly in paucibacillary leprosy in association with an active tuberculoid lesion or as part of the reactional state, but is rarely seen in multibacillary form. It develops in either peripheral nerve trunks or their cutaneous branches. It may gradually shrink, undergo calcification or produce serious complications such as increase in size or intensity of sensory loss, muscle palsies and limb deformities, that require prompt medical or surgical intervention. Deep seated abscesses or those occurring along cutaneous branches of nerve trunks are often misdiagnosed as semimembranous cysts, tendon sheath tumors, nerve tumors, lymphnodes etc. under unfamiliar settings, particularly by general physicians or surgeons, due to anatomical proximity of the different structures (Fig. 2).3

In our case also, the vascular surgeons mistook it for an anatomical proximity of the different structures (Fig. 2).3

In our case also, the vascular surgeons mistook it for external jugular vein thrombosis even in the absence of characteristic clinical signs and symptoms. As an isolated manifestation, in the absence of the classical lesion of leprosy on the cheek, it would have been difficult for us also to clinically differentiate it from other entities that present as globular swellings in the neck (Table 1).

Management of nerve abscess is governed by its impact on the affected nerve and skin. If the function is undisturbed, only antileprosy treatment is sufficient. Systemic steroids are used during reactional states. Otherwise surgical intervention like incision and drainage and nerve exploration, epineureotomy or neurectomy may be undertaken. In our patient, due to tenderness of the abscess, corticosteroids were administered before planning surgery.

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REFERENCES


Factors Predicting Morbidity and Mortality in Idiopathic Dilated Cardiomyopathy – An Inhospital Survey

Sir,

Idiopathic dilated cardiomyopathy (IDCM) is the most common variety of primary cardiomyopathy and constitutes about 25% of all patients presenting with cardiac failure. The clinical course in adults can be characterised as unpredictable with majority of deaths occurring within three years of the onset of symptoms. Parameters associated with favorable outcome include lower NYHA class, young age, and female sex whereas syncope, gallop rhythm, atrioventricular and intraventricular conduction defects (IVCD), right ventricular dilatation (RVD) carries adverse outcome.1 The prognostic significance of ventricular arrhythmias (VA) & left ventricular end-diastolic dimension (LVEDD) remain unclear. In view of insufficient data about predictors of mortality and severity of IDCM in India, this study was undertaken.

Sixty-eight cases of IDCM satisfying WHO definition were studied during their hospital stay. Electrocardiography (ECG), Holter monitoring, Doppler echocardiography and other relevant investigations were done and their clinical outcomes noted.

Out of the 68 cases studied, there were 40 males (58.9%) and 28 females (41.1%) giving a male to female ratio of 1.43. The incidence of death was significantly more in males and in older age group. The important clinical feature associated with adverse outcome in these patients was syncope. Of the 68 cases, 12 (17.64%) died during the hospital stay of which 8 (66.6%) had syncope. The association between syncope and death was significant and irrespective of the NYHA class of the patient.

Electrocardiographic study revealed that 14 cases had IVCDs in the form of LBBB, LAHB, LPHB etc. Six of these 14 cases died (42.9%). This indicates that in patients with IVCDs there is significant increased incidence of death. Six patients were having high-grade VAs (Lown grade ≥4) of which 4 (66.6%) died which was significantly high compared to the group without it. Of the total 12 cases who died in the present series, 4 (33.3%) had high-grade VAs with a mean EF of 42.5% whereas 8 cases who died without VAs had a mean EF of 34.1%. This indicates that high-grade VAs can be taken as a major independent risk factor irrespective of the pump indices. This finding is in contrast to that of a