An Uncommon Syndrome of a Common Disease

A 52-year old female presented with history of dyspnoea on exertion, dry cough, intermittent fever, dryness of mouth and dryness with irritation of the eyes for 7 months. On examination there was non-tender swelling of both the parotid glands and bilateral basal crepitations. Ophthalmic evaluation showed nebulomacular opacities in the left eye. Bilateral hilar adenopathy with bilateral lower zone reticulonodular opacities were noted on chest radiograph. Computed tomography (CT) of the chest using contrast showed bilateral hilar and subcarinal lymphadenopathy with perihilar, peribronchial thickening and scattered subpleural parenchymal nodules (Figs. 1, 2). A gallium 67-citrate scan demonstrated increased activity bilaterally in the lacrimal and parotid glands suggestive of ‘Panda’ sign (Fig. 3). Slightly increased activity was appreciated diffusely throughout the right lung. Serum angiotensin converting enzyme (ACE) level was elevated to 72 units/l (Normal 2-52 U/L). Transbronchial lung biopsy (TBLB) revealed thick walled alveoli lined by type II pneumocytes, interstitial fibrosis, mononuclear cell inflammation in the interstitial space and presence of non-caseating epithelioid cell granulomas. A diagnosis of Miculicz syndrome with stage II pulmonary sarcoidosis was made. The clinical syndrome associated with parotid swelling, fever and keratitis secondary to impaired lacrimation in absence of acute uveitis and facial palsy is known as Miculicz syndrome as was observed in our patient.

67Gallium citrate is a radioactive iron analogue that quickly binds to transferrin in the blood and carried to all tissues as a gallium-transferrin complex. In normal persons gallium is taken up by the liver (5%), spleen (1%), kidneys (2%), bone marrow (5%) and skeleton (13%). Gallium also accumulates in lacrimal glands, lymphoid tissue in the nasopharynx and the stimulated breast. At sites of inflammation, infection, active leucocytes (which contain lactoferrin) release this protein into the extracellular space when the cells are damaged. Lactoferrin has higher affinity for gallium than does transferrin especially at acid pH hence it accumulates at the inflammatory site due to binding to acid mucopolysaccharide present there. Siderophores on microorganisms also avidly bind gallium.

Focal accumulation of 67Ga in the nasopharynx, parotid gland, and lacrimal gland, distributed symmetrically, produces a distinct pattern, the ‘Panda’ sign that is strikingly similar to the dark marking of the giant panda. A distinctive intrathoracic lymph node uptake of 67Ga resembling Greek letter lambda is referred to as the ‘lambda sign. While separately lambda and panda signs are not specific for sarcoidosis, in combination with clinical features these findings represent a highly specific pattern suggestive of sarcoidosis.

Sarcoidosis is a chronic multisystem granulomatous disorder. All parts of the body can be affected, the most frequently being the lung. Involvement of the skin, eyes, lymph nodes and liver is common. The disease is often acute or subacute and self-limiting, but in many individuals it is chronic, waxing and waning for many years. The disease manifests clinically only in organs where it affects function (such as lungs and eyes) or in organs which are readily observed (such as the skin) or by X-rays (hilar nodes). Liver granulomas remain asymptomatic clinically (the only clue being raised serum alkaline phosphatase and gamma glutamyl transferase).

S Nanaware*, Dipti Gothi**, JM Joshi***
*Resident; ** Lecturer; ***Professor and Head, Department of Respiratory Medicine, Topiwala National Medical College, BYL Nair Hospital, Mumbai 400 008. Received :5.4.2004; Revised : 1.6.2004; Accepted :10.6.2004