Lumpy jaw revisited

Case

We care for an 84-year-old lady with Wegener’s granulomatosis. Ten months ago, she presented with acute renal failure and lung haemorrhage. She had bilateral patchy pulmonary infiltrates. A renal biopsy revealed rapidly progressive glomerulonephritis with >50% crescent formation. The immunofluorescence showed a pauci-immune pattern. She required haemodialysis for 3 weeks before responding to cyclophosphamide and prednisone. Aside from polymyalgia rheumatica, temporal arteritis, macular degeneration, atrial fibrillation and mitral insufficiency, she enjoyed reasonably good health. Her response to cyclophosphamide and prednisone was gratifying and her serum creatinine concentration decreased to 170 μmol/l. This admission was routine for her tenth intravenous cylcophosphamide treatment.

An incidental finding on this admission was a 2 × 1 cm and a 1.5 × 1 cm lump on her right jaw (Figure 1). She had lost a tooth on that side 2 months earlier. The mass was painless and firm. We considered the possibility that her disease involved her parotid gland. A review of Dr Wegener’s original article did not disclose such involvement [1]. Other possibilities that crossed our mind were lymphadenopathy, abscess formation, parotid gland tumours and enlargement of the juxtaoral organ of Chievitz. The juxtaoral organ is a normal permanent anatomical structure located within the soft tissue overlying the angle of the mandible in the buccotemporal space. Although the sensory organ nature of Chievitz’s organ, repeatedly mentioned in German publications, has been neglected in the last decade by American anatomists and pathologists, Ide et al. [2] recently found squamous epithelium accompanied by Pacinian corpuscles residing there. Their fortuitous finding supports a mechanosensory function for Chievitz’s organ.

Question

What do you think the lump might be?
Four weeks later, the lump had grown larger. A computerized tomographic scan was done (Figure 2). The mass was in the submandibular region. There was no bony destruction. The radiologists thought it might be a granuloma, but how they could tell remained a mystery to us. We finally, together with the patient, lost patience with the lump and called upon experts to excise the lesion. We hoped that we could pre-empt Dr Wegener and describe the first case of Wegener’s in a salivary gland. Our pathologists revelled with delight when they proved us wrong. Macroscopically, there was a cavity that contained purulent material with *Actinomyces* drusen swimming in the fluid. They provided a silver stain (Figure 3, upper panel) and an H&E stain (Figure 3, lower panel) that corroborated the initial impression.

*Actinomyces israeli* is an anaerobic normal inhabitant of the mouth, especially in the teeth and tonsils. In tissue, the organism appears grossly in tiny varicoloured (usually yellowish) granules often referred to as ‘sulfur granules’. Histologically, the granules consist of a central tangled mass of Gram-positive mycelia surrounded at the periphery by Gram-negative, club-shaped rods. The H&E stain shows the ‘ray phenomenon’ that was responsible for the initial misclassification of *Actinomyces* as a fungus: at the periphery of the granule, filaments are radially oriented and embedded in eosinophilic material (Figure 3). We treated the patient with ampicillin and sulbactam and she did well.

Actinomycosis is a masquerader. We were not brilliant diagnosticians in any event. Interestingly, the reverse image of our patient has been described. Smith and Heaton had a patient with disseminated granulomas of his lungs, skin and right thigh [3]. The initial impression was tuberculosis; however, therapy for that condition did not improve his well being. The next diagnosis was Wegener’s granulomatosis; however, immunosuppression did not help their patient. Finally, sulfur granules were identified in a subsequent biopsy.

Our patient had ‘lumpy jaw’, generally viewed as an ungulate disease. However, the disease is also common in sheep. Hoefs and Bunch investigated 3363 mandibles of wild sheep and 1028 from domesticated varieties [4]. Lumpy jaw was widespread in wild sheep of North America, but it was rare or absent in wild sheep from Eurasia. They drew evolutionary conclusions from their data. Actinomycosis is an infection that is often misdiagnosed and initially overlooked. So even if we do not have anything to add to Wegener’s paper, we still learned a lot from this lump.

**Our diagnosis**

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Conflict of interest statement. None declared.
References