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Published in: Tijdschrift voor Nucleaire Geneeskunde

IMPORTANT NOTE: You are advised to consult the publisher's version (publisher's PDF) if you wish to cite from it. Please check the document version below.

Document Version Publisher's PDF, also known as Version of record

Publication date:

Link to publication in University of Groningen/UMCG research database

Citation for published version (APA):

Dekker ,den, M. A. M., Vegt, van der, B., & Noordzij, W. (2019). Sarcoidosis of the parotid glands in Heerfordt's syndrome. *Tijdschrift voor Nucleaire Geneeskunde*, *41*(1), 2171-2172. https://www.tijdschriftvoornucleairegeneeskunde.nl/uitgaven?articlePage=8

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Download date: 12-10-2022

Sarcoidosis of the parotid glands in Heerfordt's syndrome

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Abstract

A patient with complaints of joint pain, fever, fatigue, bilateral facial paralysis, dry eyes and enlarged lymph nodes showed increased uptake on ¹⁸F-FDG PET/CT in the lymph nodes as well as intense increased uptake in the parotid glands.

Pattern of uptake in the lymph nodes was typical for sarcoidosis, but the uptake in the parotid glands was not. To confirm a possible Heerfordt's syndrome, incision biopsy of the right parotid gland was performed. This revealed non-necrotising epithelioid granulomatous infection of the sarcoid type, confirming Heerfordt's syndrome.

A 41-year old patient was referred for an ¹⁸F-FDG PET/CT because of joint pain, fever, fatigue, bilateral facial paralysis, dry eyes and swollen glands. He had been suffering from these complaints for approximately three months. Prior MRI brain and lumbar puncture showed leukoaraiosis, but no signs of tumour or infection.

18F-FDG PET/CT (figure 1) showed increased, intense uptake in multiple mediastinal and hilar lymph nodes, as well as supraclavicular and paraaortal lymph nodes. Furthermore, there was an increased, intense uptake in the parotid glands. CT scan was consistent with the 18F-FDG PET/CT, and confirmed enlarged cervical, mediastinal, hilar, and para-aortal

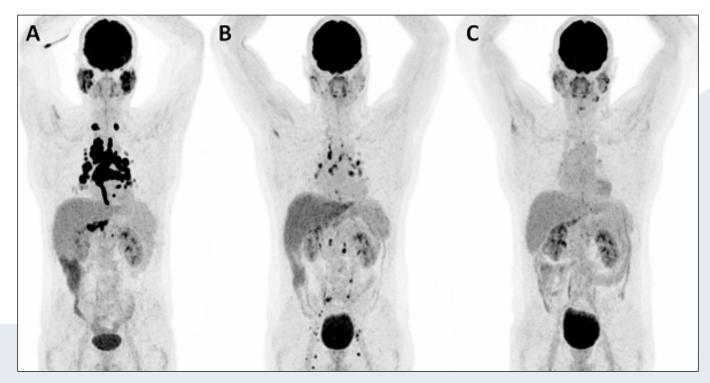


Figure 1. Anterior ¹⁸F-FDG PET maximum intensity projection, showing pathological uptake in parotid salivary glands and lymph nodes in supraclavicular, mediastinal, lung hilum and retroperitoneal stations (A). Follow-up after one year, showing normalised uptake in the parotid salivary glands and significant decrease in uptake in the lymph nodes (B). Follow-up after two years, showing a relapsed uptake in the parotid salivary glands (C).

lymph nodes, and heterogeneous enlargement of the parotid glands. The distribution and increased uptake in lymph nodes is typical for sarcoidosis. However, increased ¹⁸F-FDG uptake in the parotid glands is not a common finding. A possible explanation for increased parotic gland accumulation is Heerfordt's syndrome (1,2). However, lymphoma should also be in the differential diagnosis. This patient underwent an incision biopsy of the right parotid gland to determine the nature of the ¹⁸F-FDG accumulation. Histopathologic analysis (figure 2) revealed a non-necrotising epithelioid granulomatous infection of the sarcoid type, thereby confirming the diagnosis sarcoidosis, specifically Heerfordt's syndrome. Furthermore, the ophthalmologist was consulted and ascertained small retinal infiltrations in his left eye.

Afterwards, this patient was treated with high-dose prednisolone, resulting in a decrease in - but no normalisation of - the parotid gland and lymph node uptake. At follow-up by the ophthalmologist, retinal infiltrates had vanished completely. Two years after his initial presentation, this patient's follow-up ¹⁸F-FDG PET/CT showed an increased uptake in parotid glands and cervical lymph nodes again, indicating a flare-up of the sarcoidosis. Since he did not suffer from any complaints at that time, low-dose prednisolone was continued. Heerfordt's syndrome is a rare manifestation of sarcoidosis, and is present in approximately 0.3% of all patients presenting with sarcoidosis. Patients usually suffer from uveitis, swelling of the parotid glands and palsy of the facial nerve, on top of the clinical signs of sarcoidosis (cough, dyspnoea, chest pain, fever,

weight loss, joint pain and erythema nodosum). This rare, yet typical distribution pattern is seldom seen on ¹⁸F-FDG PET/CT.

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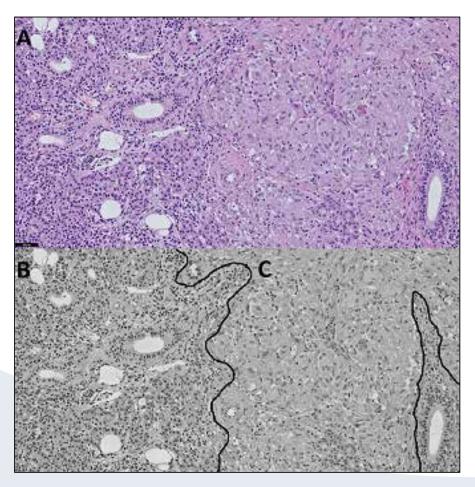


Figure 2. Microscopy image of the parotid gland biopsy (A) at 20x magnification, with normal parotid gland tissue (B) demarcated by the non-necrotising sarcoid type granuloma (C).