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Cervical metastasis masquerading as Kimura disease in a patient with nasopharyngeal carcinoma

Przerzut szyjny naśladujący chorobę Kimury u pacjentki z rakiem nosogardzieli

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Abstract

Aim: To highlight the challenges involved in the diagnosis and management of synchronous tumours. A synchronous head and neck lesion brings the attending physician into a quandary, as there is no gold standard for managing such patients. Failure to respond to treatment should provoke physicians into considering other possible diagnoses, as often a secondary lesion may be the hidden culprit. **Case report:** Herein, we present a case of a synchronous cervical mass: Kimura disease and nasopharyngeal carcinoma in a 48-year-old woman. She was initially treated for Kimura disease based on fine-needle aspiration cytology evidence, and later found out to have a synchronous malignancy in the fossa of Rosenmüller. Failure of Kimura disease to respond to steroid treatment led to a repeated examination, which resulted in the diagnosis of nasopharyngeal carcinoma. The patient was treated with concurrent chemoradiotherapy. **Conclusion:** Kimura disease, a rare benign head and neck tumour, is treated successfully with steroids. Persistent swelling coupled with treatment failure should alert physicians to repeat patient assessment, as improper disease management leads to severe complications.

Keywords: Kimura disease, nasopharyngeal carcinoma, synchronous tumour

Streszczenie

Cel: Celem pracy jest zwrócenie uwagi na wyzwania związane z diagnostyką i leczeniem nowotworów synchronicznych. Guzy synchroniczne w obrębie głowy i szyi sprawiają trudności specjalistom, ponieważ nie ma „złotego standardu" postępowania w takich przypadkach. Brak odpowiedzi na wdrożone leczenie powinien skłaniać lekarza do poszukiwania innych możliwości diagnostycznych, ponieważ ukrytą przyczyną może być zmiana wtórna. **Opis przypadku:** Przedstawiamy przypadek guzów synchronicznych umiejscowionych w obrębie szyi – guza rozpoznanego jako choroba Kimury oraz raka nosogardzieli – u 48-letniej pacjentki. U kobiety początkowo rozpoznano chorobę Kimury na podstawie wyników analizy materiału cytologicznego metodą biopsji aspiracyjnej cienkoigłowej, jednak w późniejszym okresie wykryto złośliwą zmianę synchroniczną umiejscowioną w zachyłku Rosenmüllera. Ze względu na brak widocznej odpowiedzi na leczenie steroidami u kobiety wykonano kolejne badania, na podstawie których ustalono rozpoznanie raka nosogardzieli i wdrożono jednoczesną chemioradioterapię. **Wnioski:** Chorobę Kimury – rzadko występujące schorzenie, które objawia się występowaniem niezłośliwych guzów podskórnych w obrębie głowy i szyi – można skutecznie leczyć steroidami. Utrzymujący się obrzęk wraz z niepowodzeniem wdrożonego leczenia powinien skłaniać lekarza prowadzącego do ponownej oceny pacjenta, ponieważ niewłaściwe postępowanie może skutkować poważnymi powikłaniami.

Słowa kluczowe: choroba Kimury, rak nosogardzieli, guz synchroniczny

INTRODUCTION

Synchronous head and neck tumours, albeit not uncommon, remain a conundrum to the attending physicians. The diagnosis and management of the condition are challenging, as the nature of lesions is not clearly understood, and there are, as yet, no precise treatment guidelines. Kimura disease (KD) is a benign chronic inflammatory tumour which was first described by Kimura and colleagues in 1948[1,2]. KD normally presents as deep subcutaneous swelling within the head and neck with regional lymphadenopathy as well as salivary gland involvement[3,4]. KD traditionally can be treated either by surgical excision or pharmacologically with oral corticosteroids[2,3]. Failed medical treatment should alert the physician to exclude other types of tumour or even malignancy.

CASE PRESENTATION

A 48-year-old woman with underlying hypertension presented with a two-year history of painless left neck swelling which was progressively increasing in size. The growing mass was not associated with any obstructive or constitutional symptoms. The patient denied any trauma or associated upper respiratory tract infection, and her family history was unremarkable. Besides that, there was no history of nasal or otological symptoms.

Upon examination, the patient appeared comfortable under room air. Cervical examination revealed a left neck mass occupying levels II and III, measuring 4 × 5 cm, firm, nontender, not fixed to underlying structures. The laryngeal framework was intact. Oral cavity and oropharyngeal examinations were normal. Flexible nasopharyngolaryngoscopy showed no abnormality. Nasal and ear examinations were also unremarkable. Vital signs as well as the standard blood investigations were within the normal ranges. Fine-needle aspiration cytology showed eosinophilic infiltrates against a background of lymphoid cells and fibrotic stroma, which was suggestive of KD (Fig. 1).

The patient was started on oral steroid therapy. However, the neck swelling increased in size despite completing a two-month steroid cycle. Incisional biopsy was performed,

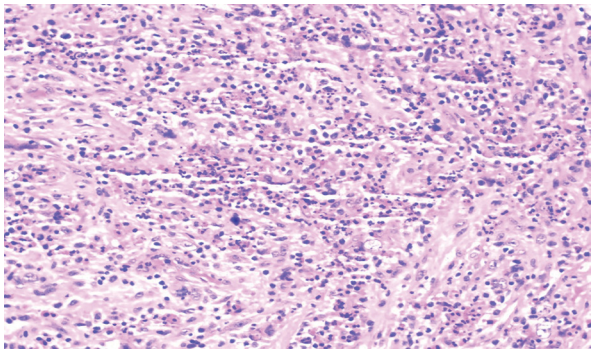

Fig. 1. FNAC revealing eosinophilic infiltrates against a background of lymphoid cells and fibrotic stroma

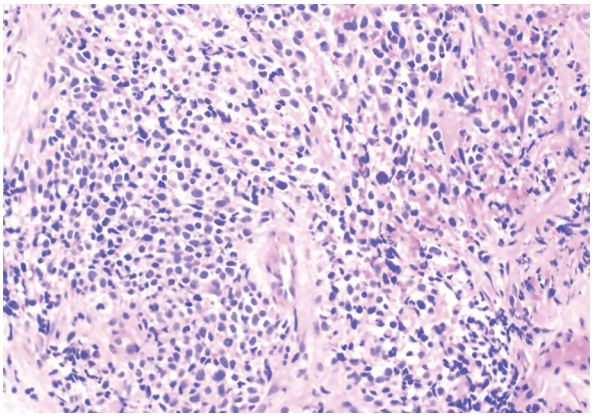

Fig. 2. HPE showing squamous epithelium infiltrated by atypical cells, exhibiting large hyperchromatic and pleomorphic cells with prominent nucleoli

as the mass was huge, and revealed metastatic carcinoma (Fig. 2). A contrast-enhanced computed tomography scan of the base of the skull to the abdomen revealed bilateral cervical and left supraclavicular necrotic lymphadenopathies with no evidence of nasopharyngeal, intrathoracic or intraabdominal lesions.

Nasoendoscopic examination was repeated after the biopsy, and to our surprise revealed a nasopharyngeal mass bilaterally obliterating the fossa of Rosenmüller. Histopathological examination revealed a non-keratinising type of squamous cell carcinoma (Fig. 3) suggestive of nasopharyngeal carcinoma (NPC) T1N2M0. The patient was then started on concurrent chemoradiotherapy.

DISCUSSION AND CONCLUSION

KD is a chronic inflammatory condition presenting as painless solitary or multiple subcutaneous nodules, asymmetric, mostly located in the head and neck region, with coexisting lymphadenopathy in 30–40% of the cases[1]. Typical areas for the nodules are preauricular, submandibular, and popliteal regions as well as the oral cavity, larynx, and parotid glands[5].

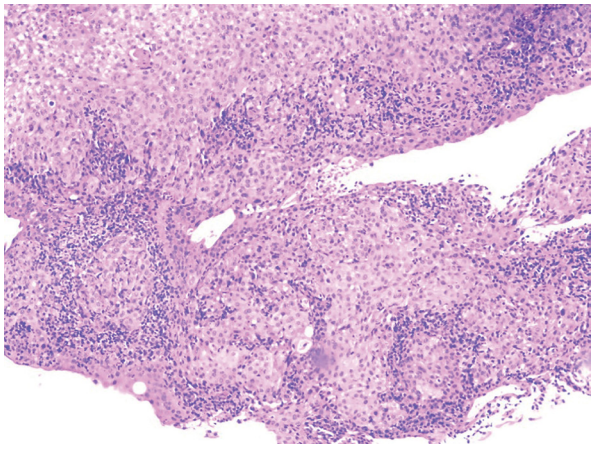

Fig. 3. HPE of fossa of Rosenmüller mass revealing keratin pearls suggestive of NPC

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They are rarely reported in other locations like the eyelids, lacrimal glands, orbits, axilla, groins, forearms, and kidneys[1,2,6–8]. On the other hand, NPC is one of the most common malignancies seen within the Asian populations, with an incidence varying between 30–80 cases per 100,000 per year[5]. The most common presentation of NPC is a painless neck mass[6]. KD has been reported to coexist with cervical lymphadenopathy in 30–40% of cases. However, a synchronous metastatic lesion is yet to be reported.

NPC has a male predominance, with a male to female ratio of 3:1[6]. The aetiology of NPC has been linked to Epstein–Barr virus infection[7], genetic susceptibility, and consumption of food containing carcinogenic nitrosamines such as salted fish whilst the aetiology of KD is still not entirely clear[1,8].

The most common presentation of NPC is cervical lymphadenopathy, either unilateral or bilateral, though the condition can also present with unilateral ear complaints such as conductive hearing loss, middle ear effusion, and tinnitus[6,9]. Other possible manifestations, including headache and nasal symptoms such as nasal congestion and epistaxis[5], are not uncommon, either. As in our patient, she presented with bilateral painless neck swelling which fits both KD and NPC. In a patient with the sole presentation in the lymph nodes, fine-needle aspiration cytology (FNAC) is the cornerstone. If a mass is present in the fossa of Rosenmüller, a biopsy of the primary lesion is confirmative.

Histopathological examination (HPE) is the gold standard in diagnosing NPC[10]. According to the World Health Organization, histologically NPC is classified into three subtypes: type I – keratinizing squamous cell carcinoma, type IIa – non-keratinizing carcinoma, and type IIb – undifferentiated carcinoma. Subtype IIb is the most common, and it responds well to local therapy but has a higher rate of metastasis[9].

On the other hand, KD is suspected when key microscopic findings of marked lymphoid hyperplasia with eosinophilic infiltration are present[11]. It is noteworthy that angiolymphoid hyperplasia with eosinophilia (ALHE) may show similar microscopic features to KD, but "histiocytoid" and "epithelioid" blood vessels seen in ALHE are lacking in KD[11]. Hence, FNAC alone is insufficient to diagnose KD, and tissue biopsy is required for HPE to confirm the diagnosis[1,3,11]. Other tests that can support the diagnostic work-up of KD include peripheral blood eosinophils and IgE level[1,4,11]. These tests however, were not done in our patient.

Magnetic resonance imaging (MRI) is known for its soft tissue sensitivity and regarded as a preferred imaging modality to detect primary disease as well as submucosal lesion which may be missed on nasoendoscopic procedures[7,12]. A recent study on MRI and endoscopy in 246 cases with suspicions of NPC showed an improvement in sensitivity to detect primary pathology (100% vs. 88%, $p = 0.003$)[12]. Computed tomography (CT) has a low sensitivity to diagnose primary pathology and, as a consequence, is not a preferred imaging option[10]. Nevertheless, due to time, cost constraints as well as limited availability, CT is the most commonly used option.

KD can be treated either surgically or medically. We opted for pharmacological treatment in our patient and started therapy with oral steroids. Besides that, surgical excision can be undertaken, where possible. Persistent swelling despite oral steroids led to a repeat biopsy, which resulted in the diagnosis of NPC. The patient was given neoadjuvant chemotherapy followed by radiotherapy (RT), according to the guidelines of the American Joint Committee on Cancer (AJCC) 2017, which are known to improve progression-free survival and overall survival[7,13]. As for the coexistence with KD, RT has been proven to treat KD successfully[14].

Synchronous lesions within the head and neck are not unheard of. Knowledge of this entity along with meticulous history-taking and patient examination are necessary. Failure of treatment should alert the attending physician to undertake further examinations, as a delay of appropriate treatment may lead to devastating complications.

Conflict of interest

All authors have no conflict of interest.

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