# A mucoepidermoid carcinoma in a young man with intellectual disability: review of oral cancer in people with intellectual disability

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Oral tumors in patients with intellectual disabilities (ID) remain poorly documented, despite cancer incidence suggesting that malignancies are globally as frequent in this group as in the general population. A clinical case of a 36-year-old man with severe ID presenting with a mucoepidermoid carcinoma of intermediate grade in the right mandible is reported. Delayed diagnosis and problems managing complementary chemotherapy and radiotherapy are described. The literature review reported only 27 cases of malignant tumors in patients with ID. This finding indicates that oral tumors in patients with ID may be less frequent than in the general population, are usually diagnosed at an advanced stage, and may occur in patients who are younger than the general population. Diagnosis and treatment are difficult, implying a comprehensive knowledge of the underlying condition of each individual and the need for good communication skills to obtain patient cooperation, including an understanding of how the patient expresses pain. (Oral Surg Oral Med Oral Pathol Oral Radiol 2013;115:e22-e27)

In addition to the heavy burden of mental impairment, people with intellectual disability (PWIDs) may suffer from cancer.[1] Two recent epidemiologic studies on cancer incidence suggest that globally malignancies are as frequent in PWIDs as they are in the general population.[2,3] These tumors remain poorly understood and, in particular, the relative importance of the environmental and constitutional risk factors on oral tumors in this population is not known. Difficulties in communicating pain or discomfort, conditions for access to medical centers, ability to cooperate during care procedures, and the ability of care-givers to perform routine hygiene are key factors for oral health in PWIDs. Poor oral health is a complication of disability in children, and their oral health status worsens with age.[4] Among individuals with intellectual disabilities (ID), those who are less cooperative for complete oral examination and hygiene or care procedures are at higher risk of experiencing delayed diagnosis of oral tumors. A recent review of cancer and ID drew attention to "the lack of information on intellectual disability and oral cancer."[5] Following a letter to the Editor drawing attention to this subject,[6] we present a literature review of oral cancer in PWIDs, including a case report of a mucoepidermoid carcinoma of the mandible in a young man with post-encephalitis ID. We hope this text will provide a basis for clinical monitoring and further research on the subject.

## CASE REPORT

A 36-year-old man with severe ID, living in an institution, was referred for a lump on the right posterior region of the mandible. He was the issue of a normal pregnancy and was in

## Statement of Clinical Relevance

Oral tumors in people with intellectual disability are rare and usually are found at an advanced stage. Delays in diagnosis could be improved by educating clinicians, by improving methods for communication, and potentially by performing periodic radiographic imaging.

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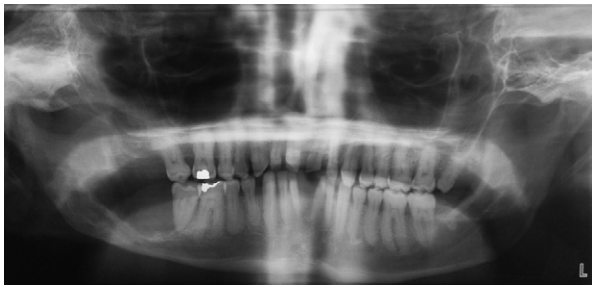

Fig. 1. Panoramic radiograph undertaken for the reported case before the first general anesthesia indicated for dental extraction and tumor resection. A wide lytic lesion is observed behind the lower right second molar. Periradicular bone deposition is seen in the adjacent teeth.

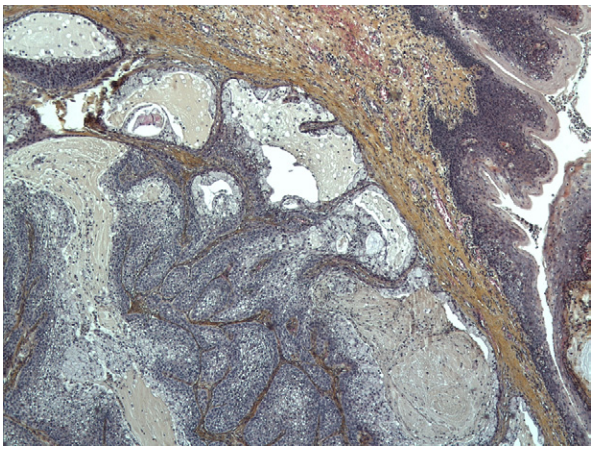

Fig. 2. Hematoxylin–eosin–safran stain × 100. Histologic aspect of the tumor at the first surgical resection.

good health during infancy. At 18 months of age, he developed varicella encephalitis, which led to ID. At 6 years of age, he developed herpetic encephalitis, resulting in epilepsy and a regressive left hemiplegia. A lumbar scoliosis was surgically corrected during childhood. His parents were in good physical and mental health but a brother, born 16 years before him, suffered from mild ID of unknown cause. Another brother, born 8 years before him, suffered from autism and ID. His third brother and his sister have had normal intellectual development. There is no particular cancer history in his parents. A second cousin was operated on at 18 years of age, 3 years before him, for a Ewing sarcoma of the left mandible. After partial mandibular resection, she underwent chemotherapy and radiotherapy according to the Euro-Ewing 99 protocol. She is currently in remission.

As is usual for individuals in the general population, the patient had had annual dental check-up at a dental clinic and regular scaling since March 1997. He had no regular dental radiographs. He was described as poorly cooperative for dental examination and hygiene sessions. In October 2009, during his annual examination, motility of the lower right second molar was observed. Six months later, he was treated for localized gingivitis around this tooth. Local treatment was repeated 1 month later because of persistent gingivitis and the appearance of a lump. At the end of this period, he received antibiotics for a dental abscess. Treatment was changed after 1 week because of lack of effectiveness. A panoramic radiograph showed a large lytic area of nearly 4 cm in greatest diameter behind the lower right second molar. There were also condensing osteitis-like changes observed in the adjacent teeth (Figure 1). The second molar extraction and tumor resection were performed under general anesthesia. During 3 weeks following resection, the gum remained inflamed, with preauricular swelling, and the patient refused to eat. The surgical resection measured 4.5 × 3.8 × 2.5 cm and was largely occupied by a central mucoepidermoid carcinoma of intermediate grade (Figure 2). The proliferation index was low, at 5%, and there was less than 1 mitosis per 10 high-power fields. Postoperatively, the patient was treated for anemia and had a transfusion. The diagnosis of jaw carcinoma was confirmed 1 month after surgery. A computed tomography scan under general anesthesia was performed for complete anatomical examination and a search of possible metastases and preparation for surgery on the jaw. Metastasis was not detected. Four months after the first surgery, a right mandibular resection with homolateral cervical lymph node dissection was performed. There was no grossly residual tumor. The solid and cystic tumor measured 11 cm in its anterior–posterior diameter. Few tumor cells in soft tissues reached the surgical anterior margin section. Other bone and soft tissue sections were free of tumor cells. One of 8 lymph nodes contained 3 micrometastases. Given the severe ID of this patient, complementary radiotherapy and chemotherapy were not administered. During the postoperative period, he developed a local infection and a large fistula, which was closed 5 weeks later. Because of these local complications and his mental state, he was fed through a gastric tube.

The patient returned to his institution and tolerated the gastric tube well. There was no weight loss and he seemed to accept the situation as long as the pain was controlled. Eight months after surgery, he appeared with a local recurrence, revealed by right mandibular swelling and bleeding. Bleeding was controlled by local hemostatic dressings. Given the extent of the tumor at recurrence and the patient's mental condition, no additional treatment was proposed. During the course of the disease, the illness and the proposed treatment were explained to the patient as far as he could understand. During his hospitalization, the hospital nursing team was advised by members of the residential care staff about his behavior and symptoms to allow optimal communication and cooperation with the patient. The San Salvador pain assessment scale[7] was used and adapted for him because he could not make the usual facial movements as a result of his tumor and the surgical treatment. Despite his limited understanding, the patient remained cooperative during the course of his disease. He died 20 months after the first discovery of his tumor.

## REVIEW OF THE LITERATURE

The search of the literature was conducted on PubMed using the words "cancer," "carcinoma," "sarcoma,"