

AN UNUSUAL CASE OF LINGUAL ALVEOLAR SOFT PART SARCOMA DURING PREGNANCY

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SUMMARY

Objective: We report a case of alveolar soft part sarcoma (ASPS) of the tongue that presented in the third trimester of pregnancy.

Case Report: An 18-year-old, gravida 1, para 0, woman with ASPS of the tongue in the 31st week of pregnancy was described. Approximately 4 months after her first diagnosis, she was referred to our clinic for the delivery of her baby and additional treatment. At 32 weeks of gestation, a healthy male infant weighing 2,220 g was delivered by cesarean section. After an uneventful postoperative course, she was referred to the department of otorhinolaryngology and maxillofacial surgery for further investigation and treatment.

Conclusion: ASPS of the tongue presenting in pregnancy is an extremely rare combination, and to our knowledge, this is the first reported case in the English literature. The diagnostic and therapeutic management of the pregnant patient with cancer, like ASPS, is especially difficult, because it involves both the mother and fetus. [*Taiwan J Obstet Gynecol* 2008;47(2):212–214]

Key Words: alveolar soft part sarcoma, cancer, pregnancy, tongue

Alveolar soft part sarcoma (ASPS) is a very rare malignant mesodermal tumor of unknown origin, representing 0.5–1% of all soft tissue sarcomas [1]. The lesion almost always presents in the muscles of the extremities, and only a few cases in the oral and perioral regions have been reported [2–4]. To our knowledge, there are no published reports of ASPS of the tongue during pregnancy. Here, we report a case of ASPS of the tongue that presented in the third trimester of pregnancy.

An 18-year-old, gravida 1, para 0, woman with ASPS of the tongue in the 31st week of pregnancy was examined on August 9, 2006. Her medical and family histories were unremarkable. She had never smoked and did not use alcohol. The patient had noticed swelling on the dorsal portion of her tongue and discomfort and pain in her throat 5 months before being referred to our clinic. She visited an otolaryngologist with her complaints. Magnetic resonance imaging scan was performed, and

the sagittal scan revealed a 4-cm sized, well-defined hyperintense signal mass involving a large area on the dorsal side of the tongue extending into the oropharynx. An excisional biopsy was performed, and the histopathologic diagnosis of ASPS was made. Then, she received one cycle of chemotherapy with a combination of ifosfamide (2 g/m²), mesna (at 20% of the ifosfamide dose, given at 0, 4 and 8 hours as an intravenous bolus) and adriamycin (20 mg/m² intravenously). During this period, her pregnancy continued normally. The patient was closely followed; her lesions increased gradually in size and her complaints increased. Approximately 4 months after the first diagnosis, she was referred to our clinic for her delivery.

Examination revealed an ulcerated lesion, measuring approximately 45 × 30 mm on the right dorsum of the tongue, with surrounding induration. Tongue mobility was limited to forward protrusion. There were many palpable cervical lymph nodes on the bilateral sides (Figure). Other examinations and laboratory findings were within normal limits. The patient's general condition gradually worsened, which was related to her limited breathing. Therefore, she underwent tracheostomy by an otolaryngologist.



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Figure. Frontal view of the patient.

Because of her worsening condition, our hospital staff proposed cesarean section. After an extensive discussion with the patient and her husband, delivery of the pregnancy was accepted by the couple. In the 32nd gestational week, a healthy male infant (birth weight, 2,220 g) was delivered by cesarean section under epidural anesthesia. The postoperative course was uneventful. Then, she was referred to the department of otorhinolaryngology and maxillofacial surgery for further investigation and treatment.

Cancer complicating pregnancy is a rare coexistence, and the estimated incidence in developed countries is 1 in 1,000 pregnancies. Cancer diagnosed during pregnancy is a dramatic event, with profound effect on the lives of the patient, children, family and physician. This gives rise to opposing emotional reactions in these women; they are happy with their pregnancy but are usually devastated when they hear that they have cancer. The most common malignancies associated with pregnancy include cervical cancer, breast cancer, melanoma, lymphomas, and leukemias [5]. Oral cancer during pregnancy has been described, though it accounts for less than 2% of all cancers [6]. To our knowledge, there are no published reports in the English literature of ASPS of the tongue during pregnancy.

ASPS was first described as an entity and classified in 1952 by Christopherson and colleagues [7]. The clinical and histologic appearance of this tumor can be misleading, and clinical characteristics include slow growth and a high risk of mortality. Local recurrence is rare, but metastasis to the lung, bone and brain may occur. ASPS rarely arises in the oral cavity; but when it occurs, it most often originates from the tongue [8]. The treatment choice is generally surgical excision with sufficient margins. Surgical approach with chemotherapy or radiotherapy is useful in some patients. In our patient, although chemotherapy with a combination of ifosfamide, mesna and adriamycin had been given after the

excisional biopsy, the treatment modality did not seem to be adequate because of her pregnancy.

The physiologic changes of pregnancy are all potentially capable of promoting neoplastic growth. The major reason for suspecting that pregnancy adversely affects the clinical course of cancer is the immunologic tolerance that characterizes both conditions. The immune system is suppressed during the first 20 weeks of gestation, which in some way predisposes the subject to malignancy, modifies the biologic behavior of the neoplasm, and adversely affects the clinical course of the malignancy [9]. It has been shown that high levels of estrogen and progesterone during pregnancy may stimulate latent breast carcinoma to an active state [10]. However, these hormonal receptors are generally not found in head and neck tissues, as was the case in our patient, but the rapid progression of the disease in our case may be explained by the abovementioned pregnancy-related physiologic changes. In summary, humoral factors might act through progesterone and estrogen receptors, and the immunotolerance characteristic of pregnancy might also contribute to tumor progression.

The management of a pregnant mother with cancer is associated with considerable uncertainty concerning the timing of treatment and fears that the pregnancy may accelerate cancer growth, and is difficult and emotionally driven, often requiring the participation of the doctor, patient and the family. The main concerns of both the obstetrician and the oncologist are to save the mother's life and to protect the fetus. Optimal cancer therapy, be it chemotherapy, radiotherapy or surgery, may impose a great risk on the fetus [5,9]. A major problem is the assessment of the optimal time of delivery, as was the case for our patient. The physician has to balance the risk and the health of the pregnant patient by delaying therapy against the risk to the fetus' health by beginning the potentially teratogenic chemotherapy and/or radiotherapy. As a result, optimal therapy for the tragic diagnosis of cancer in pregnancy requires a collaborative and interdisciplinary approach between gynecologists, surgeons, oncologists, obstetricians, neonatologists, psychologists and other disciplines.

In conclusion, ASPS of the tongue presenting in pregnancy is an extremely rare case, and to our knowledge, this is the first reported case in the English literature. The diagnostic and therapeutic management of the pregnant patient with cancer, like ASPS, is especially difficult, because it involves both the mother and her fetus. Conflict between maternal therapy and fetal well-being is frequently observed in cases of pregnant women with cancer, and a multidisciplinary approach is crucial to the optimal treatment of the mother and her fetus.

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