Olgu Sunumu

A Case of Alveolar Soft-part Sarcoma with Cerebral Metastases

Çağatay ÖZDÖL, Hilmi Önder OKAY, Emre Cemal GÖKÇE, Ergun DAĞLIOĞLU, Ali DALGIÇ, Ahmet Deniz BELEN

Ankara Numune Eğitim ve Araştırma Hastanesi 2. Beyin Cerrahi Kliniği, Ankara

✔ Alveolar soft part sarcoma is a rare soft tissue tumor primarily within the skeletal muscles or musculofacial plane in young adults. Lung and brain are well known regions for late metastasis. Treatment modalities are surgery, radiotherapy and chemotherapy. Combination of surgery with radiotheraphy and chemotheraphy may prolong the mean survival time. Here we report a 32-year-old man presenting with an occipital mass. The patient was previously operated for masses on neck and left thigh with the same diagnosis. Histopathological specimens confirmed the diagnosis of alveolar soft part sarcoma metastatis. Despite systemic dissemination alveolar soft part sarcoma has a better prognosis if removed completely.

Key words: Alveolar soft part sarcoma, cerebral metastasis, treatment

J Nervous Sys Surgery 2009; 2(3):152-154

Beyin Metastazı Olan Alveolar Soft-part Sarkom Olgusu

✔ Alveolar soft part sarkom genç erişkinlerde nadir görülen primer olarak iskelet kasları ve muskulofasial planda izlenen yumuşak doku tümörüdür. Akciğer ve beyin iyi bilinen geç metastaz yerleridir. Tedavi yaklaşımları cerrahi tedavi, radyoterapi ve kemoterapidir. Cerrahi tedavinin radyoterapi ve kemoterapi ile kombinasyonu ortalama sağkalım süresini uzatır. Bu yazıda 32 yaşında oksipital bölgede intraserebral kitle ile ortaya çıkan genç erkek olguyu sunuyoruz. Hasta daha önce boyun ve sol kalçadan yumuşak doku kitleleri nedeniyle opere edilip aynı tanıyı almış. Histopatolojik inceleme alveolar soft part sarkom tanısı ile uyumlu gelmiştir. Sistemik yayılımına rağmen alveolar solf part sarkom tümörleri tamamen çıkarıldığında iyi prognoza sahiptir.

Anahtar kelimeler: Alveolar soft part sarkom, beyin metastazı, tedavi

J Nervous Sys Surgery 2009; 2(3):152-154

Alveolar soft part sarcoma (ASPS) is a rare soft tissue neoplasm accounting for 1 % of the sarcoma subtypes [(1)]. Lower extremity of young adults is the most common region for this tumor with characteristic histopathological findings. The mean survival time of ASPS is relatively longer than other sarcoma subtypes and its typical feature is to metastasize to lung, brain and bone after long time intervals as much as 33 years [(2)]. Another interesting characteristic of this type of sarcoma is its predisposition to metastasize to brain after or with pulmonary metastasis [(3)]. Treatment modalities including surgery, radiotherapy and chemotherapy may significantly prolong the survival time. In this report, we present a case of ASPS primarily originating from left thigh with cranial and pulmonary metastasis after 16 year interval and successful surgical management of cranial metastasis as well as radiotherapy and chemotherapy for pulmonary metastasis.

CASE REPORT

A 32-year-old man admitted to our service with complaints of severe headache and blurring of vision which started about a month ago. Systemic examination was normal except for the scar inci-

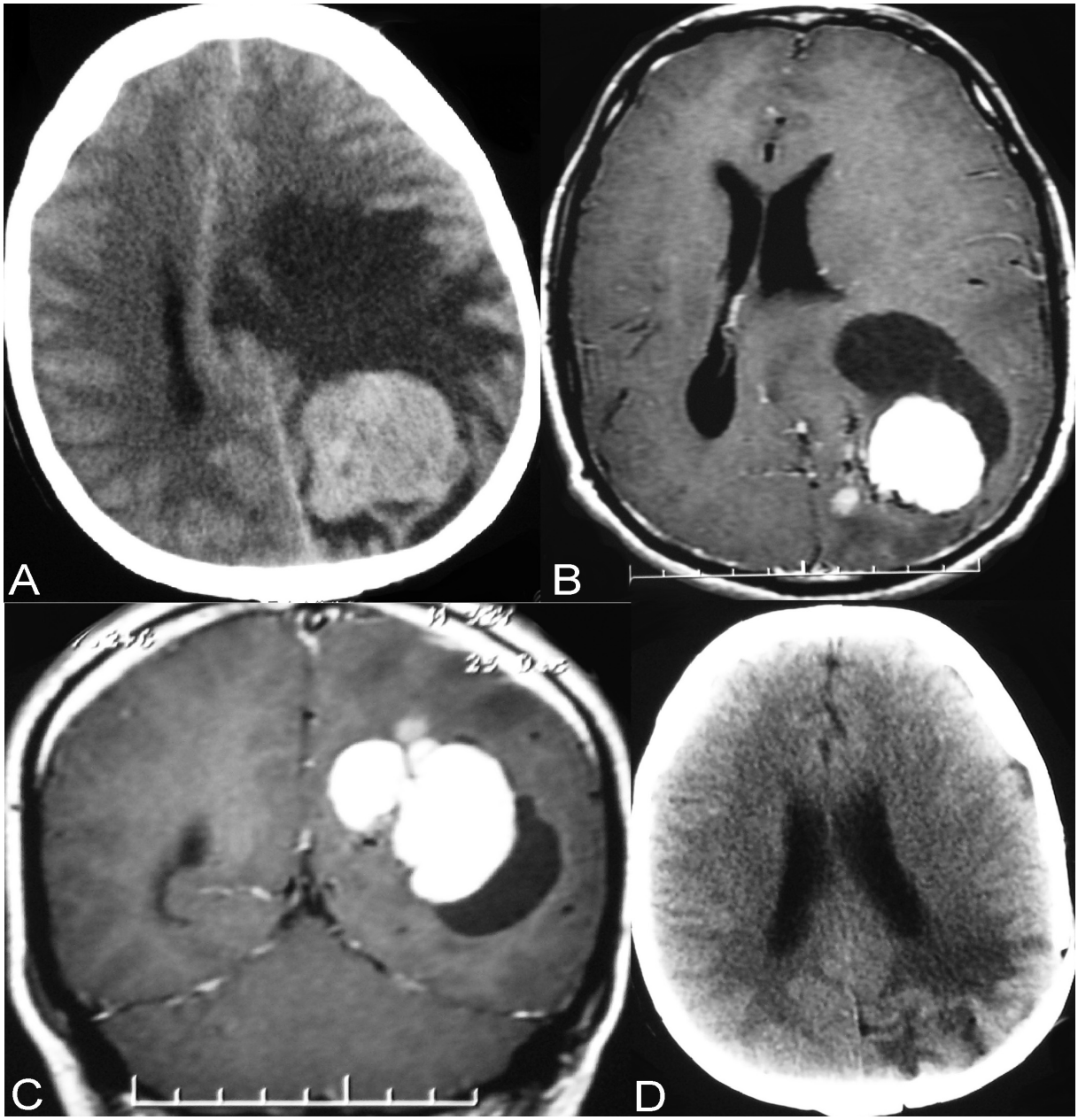

Figure 1. Preoperative contrast enhanced axial CT scan of the cranium (a) showed markedly enhanced tumor with irregular borders. Axial (b) and coronal (c) MR scans demonstrated a cystic component adjacent to the inferolateral portion of the tumor. Postoperative axial CT scan (d) demonstrates complete removal of the cystic mass.

sion on the left thigh and neck. The patient was disoriented and had severe papilledema on neurological examination. Cranial computerized tomography (CT) showed an edematous lesion in the left occipital lobe which is 4x5 cm in size (Figure 1a). Magnetic resonance imaging (MRI) revealed a markedly enhanced lesion in left occipital lobe which is 60x62x42 mm in size with a cystic component and severe edema (Figures 1b and 1c). Preliminary diagnosis was meningioma. The patient underwent surgery and heavily vascular lesion was removed completely with a left parietooccipital craniotomy. Histopathological examination of the mass was consistent with alveolar soft part sarcoma metastasis. The tumor brain interface was noted to be well demarcated on histopathological specimens and there wasn't any involvement of dural layer

and cranium. Postoperative course was uneventful without any new neurological deficit. Abdominal ultrasonography was normal which was done postoperatively. Thorax CT examination revealed multiple metastatic nodules in both lungs, the biggest of which was 3 cm. Postoperative CT confirmed total excision of the tumor (Figure 1d). Radiotherapy and chemotherapy was started for pulmonary metastasis and the patient was clinically asymptomatic at his 6 months' of follow up.

DISCUSSION

Sarcomas form 3 % of metastatic brain tumors [4]. Although ASPS is a rare type of sarcoma with an incidence of 1 % among all soft tissue sarcomas, they form one third of all metastatic soft tissue sarcomas (33 %) [5]. Metastatic sarcomatous brain lesions appear to occur in association with or following lung metastasis. In a report from Memorial Sloan Kettering Cancer Center only 4 patients among 102 patients had solitary brain lesions so that when a patient presented with neurological symptoms and history of ASPS, brain and thorax imaging is accurately indicated in the first step of evaluating the patient. The presence of lung metastases is not a contraindication to surgery in patients with alveolar soft part sarcoma [6]. Surgery is the goal and complete microscopic resection is critical in the treatment of metastatic ASPS. The mean survival time of patients who are in a good medical status, with a Karnofsky performance score >70 and who had total excision, is relatively long and are associated with a favorable prognosis. The reason of favorable prognosis with the aggressive resection of the metastatic lesions can be explained by the slow growing and indolent nature of this disease. Although the chromosome rearrangements at 17q25 and Xp11.2 in ASPS have been disclosed, the cause of prolonged survival of ASPS is unknown [7].

In the literature 5 year overall survival was reported as 83 % [8]. In our reported case, the patient was alive for more than 16 years after diagnosis.

Combination of surgery with radiotherapy and chemotherapy may prolong the mean survival time. There are some reports regarding excision of metastases provide a good prognosis all alone without giving radiotherapy or chemotherapy. In the 30-year experience with 20 patients, Katyon et al could not show a benefit from chemotherapy or radiation [8]. In conclusion, ASPS is a rare soft tissue tumor and with an adequate clinical context, it permits preoperative diagnosis. Surgical resection of solitary brain metastasis is usually effective for treatment.

REFERENCES

1. **Akiyama Y, Baba T, Ibayashi Y, Asai Y, Houkin K.** Alveolar soft part sarcoma in brain with cardiac metastasis: A case report. Int J Cardiol 2007; 114:93-5.
2. **Lillehei KO, Kleinschmidt-DeMasters B, Mitchell DH, Spector E, Kruse CA.** Alveolar soft part sarcoma: an unusually long interval between presentation and brain metastasis. Hum Pathol 1993; 24:1030-4.
3. **Lieberman PH, Brennan MF, Kimmel M, Erlandson RA, Garin-Chesa BY, Flehinger P.** Alveolar soft part sarcoma a clinicopathologic study of half a century. Cancer 1989; 63:1-13.
4. **Ogose A, Morita T, Hotta T, Kobayashi H, Otsuka H, Hirata S, Yoshida Y.** Brain metastases in musculoskeletal sarcomas. Japanese Journal of Clinical Oncology 1999; 29:245-7.
5. **Perry JR, Bilbao JM.** Metastatic alveolar soft part sarcoma presenting as a dural-based cerebral mass. Neurosurgery 1994; 34:168-70.
6. **Bindal RK, Sawaya RE, Leavens ME, Taylor SH, Guinee VF.** Sarcoma metastatic to the brain: results of surgical treatment. Neurosurgery 1994; 35:185-91.
7. **Portera CA Jr, Ho V, Patel SR, Hunt KK, Feig BW, Respondek PM, et al.** Alveolar soft part sarcoma: Clinical course and patterns of metastasis in 70 patients treated at a single institution. Cancer 2001; 91:585-91.
8. **Kayton ML, Meyers P, Wexler LH, Gerald WL, LaQuaglia MP.** Clinical presentation, treatment, and outcome of alveolar soft part sarcoma in children, adolescents, and young adults. J Pediatr Surg 2006; 41:187-93.