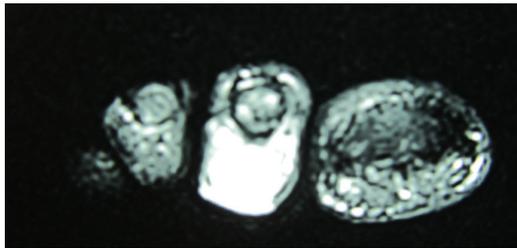
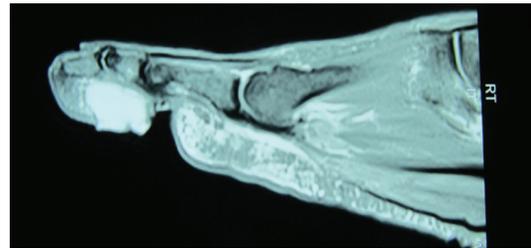




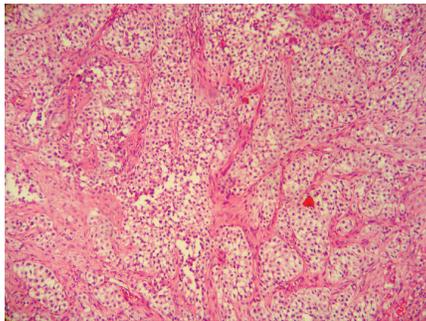
**Figure 1.** Ulceration of the tumor's overlying the skin of the second toe for 4 months duration.



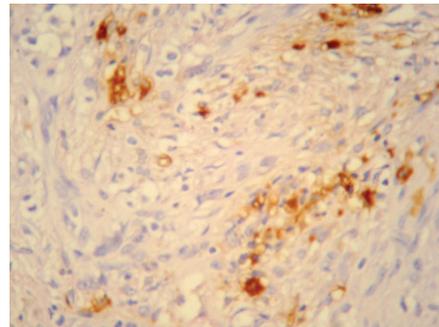
**Figure 2.** Axial MRI shows the tumor and its depth of invasion.



**Figure 3.** Sagittal MRI shows the tumor and its depth of invasion across the second phalanx.



**Figure 4.** Histopathologic findings revealed that fibrous tissue septa had divided the tumoral cells into well-defined nests and groups of pale-staining cytoplasm as well as short fascicles arrangement and a few multinucleated giant cells. The cells had vesicular nuclei and prominent basophilic nucleoli (Hematoxylin and Eosin, 40×).



**Figure 5.** Positive immunohistochemistry staining for S100.

Cite this article as: Afshar A, Ilkhanizadeh B. Photoclinic. *Arch Iran Med.* 2014; **17**(12): 853 – 854.

A 32-year-old woman was referred because of a painful recalcitrant ulcer on the plantar surface of the right second toe of 4 months duration (Figure 1). She had noticed tenderness and a palpable nodule since one year ago. The mass became larger and the overlying skin became ulcerated since 4 months ago. There was no bony lesion on X-ray. Sonography revealed a 15×11 mm soft tissue mass. Magnetic resonance imaging revealed a soft tissue mass that had occupied the entire volume of the plantar aspect

of the second toe. The tumor was located adjacent to the flexor tendons (Figures 2 and 3). There was a palpable lymph node in the right groin. Incisional biopsy was performed. Histopathologic findings revealed that fibrous tissue septa had divided the tumoral cells into well-defined nests and groups of pale-staining cytoplasm as well as short fascicles arrangement and a few multinucleated giant cells. The cells had vesicular nuclei and prominent basophilic nucleoli. Some mitotic figures were seen (Figure 4). Immunohistochemistry staining for S100 was strongly positive but HMB45 staining was negative (Figure 5). Further work-up and PET scan revealed the presence of multiple lung metastases.

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Accepted for publication: 12 November 2014

**What is your diagnosis?  
See the next page for your diagnosis**

The histopathology diagnosis was consistent with Clear Cell Sarcoma (CCS). The second toe was amputated through the second metatarsophalangeal joint. The patient was referred to an oncologist for further treatment.

CCS is a malignant ectoblast tumor that originates from the latent melanin-producing cells that had wandered from the neural crest in the embryonic period.<sup>1-7</sup> Since melanin or melanosomes have been found in the majority of CCS cases, Enzinger believed that malignant melanoma and CCS have the same origin and coined the term “malignant melanoma of soft parts” for CCS.<sup>3,5,7</sup> CCS has a clear histopathologic morphology. However, sometimes it may be difficult to differentiate the tumor from amelanotic malignant melanoma. In this situation, cytogenetic analysis helps to differentiate the tumors because the two tumors are cytogenetically different. The primary feature of CCS is the translocation of t(12;22)(q13;q13), while this translocation is not observed in malignant melanoma.<sup>1,6</sup> Positive S100 and HMB45 Immunohistochemistry are specific for the cells derived from the neural crest and malignant melanoma. In the majority of CCS cases, HMB45 and S100 immunostaining are positive. However, cases have been reported which were positive for S100 and negative for HMB45 and vice versa.<sup>1,5</sup> The histological differential diagnosis of CCS includes synovial sarcoma, fibrosarcoma, malignant Schwannoma and malignant melanoma.<sup>5</sup>

CCS is a very rare sarcoma with an incidence of 1% of all total melanomas.<sup>1,5,6</sup> CCS mainly involves the lower extremity; particularly, the tumor has a predilection for tendons and aponeuroses

of the foot and ankle regions. CCS has a very poor prognosis. Metastases are commonly present at the time of diagnosis.<sup>5</sup>

In the current case, presence of an ulcerated skin as well as the large size of the tumor relative to the size of the second toe and involvement of the deep layers of soft tissues signified poor prognosis.<sup>2</sup> In the current case, lung metastases were discovered at the time of diagnosis. We plan to continue follow up of the patients.

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