

Case report

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Solitary Plasmacytoma of the Parotid Gland with Crystalline Inclusions: A Case Report

Rani Kanthan* and Bahman Torkian

Address: Department of Pathology, Room 2868, G Wing, Royal University Hospital, Saskatoon, Saskatchewan, S7N 0W8, CANADA

Email: Rani Kanthan* - inarkanth@shaw.ca; Bahman Torkian - torkian@yahoo.com

* Corresponding author

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Abstract

Background: Solitary extramedullary plasmacytoma of the parotid gland is a rare condition. Intracytoplasmic Crystalline inclusions in the tumor are even rarer and have been reported only once in the parotid gland.

Case presentation: We report here, a case of plasmacytoma of the parotid gland with intracellular crystalline inclusions in a 73-year-old woman

Conclusion: Solitary extramedullary plasmacytoma of the parotid gland and crystalline inclusions in the tumor is of rare occurrence. The importance of such a finding with relation to tumor progression, clinical course of the disease or prognosis in general remains to be understood.

Introduction

Plasmacytoma is traditionally divided into medullary and extramedullary type, which in turn could be either solitary or multiple in distribution. The most common form of plasmacytoma is the generalized medullary form that is known as multiple myeloma. Solitary primary extramedullary plasmacytoma is a neoplasm of the plasma cells arising in regions other than bone marrow in patients with no clinical or biochemical evidence of multiple myeloma. Booth et al [1] in their review of 250 reported cases stated that primary extramedullary plasmacytoma usually affect the head and neck area. Involvement of the parotid gland is extremely rare and since the first case in 1965 [2] there are only a few documented cases in the published literature [2-15] as listed in Table 1. Although review of the literature shows a favorable prognosis, clinical behavior of this disease is not well explained. The purpose of this report is to add another case of this kind, not only because

of its rarity, but also because of its rare and interesting finding of intracellular crystalline inclusions.

Case Presentation

A 73-year-old Caucasian female was investigated at the Royal University Hospital of Saskatoon, Canada, for a non-tender mobile mass at the angle of the right mandible with a history of recent rapid growth over the past two months. Clinical examination was otherwise non-contributory. Past medical history included psoriasis, hypertension and osteoarthritis of both hip joints and treated Hodgkin's lymphoma 10 years back. Cytology of the fine needle aspiration of the parotid mass showed numerous polymorphic lymphocytes (Figure 1). Subsequently a superficial parotidectomy with conservation of the facial nerve was performed. Histopathological examination of the excised mass at initial frozen section analysis revealed the parotid gland to be completely replaced by sheets of

Table 1: Review-published Cases of Parotid Plasmacytomas

Author	Sex/Age	Treatment
1-Ustun MO <i>et al.</i> [15]	F/77	S+R
2-Hari CK, Roblin DG. [14]	F/77	S
3-Gonzalez-Garcia J <i>et al.</i> [13]	M/63	S+R
4-El-Naggar AK [4]	M/73	S
5-Kerr PD, Dort JC [12]	F/73	S+R
6-Rothfield RE <i>et al.</i> [10]	M/53	S
7-Simi U <i>et al.</i> [9]	M/58	S
8-Scholl P, Jafek BW [8]	F/60	S
9-Ebbers J [7]	M/68	R
10-Kanoh <i>et al.</i> [6]	F/78	S+R
11-Edney JA <i>et al.</i> [5]	M/38	S
12-Ferlito A <i>et al.</i> [4]	M/47	S+R+Ch
13-Pahor AL. [3]	F/61 M/69	S+Ch R
14-Vainio-Mattila J [2]	F/74	S+R

S = Surgery R = Radio therapy Ch = Chemo therapy

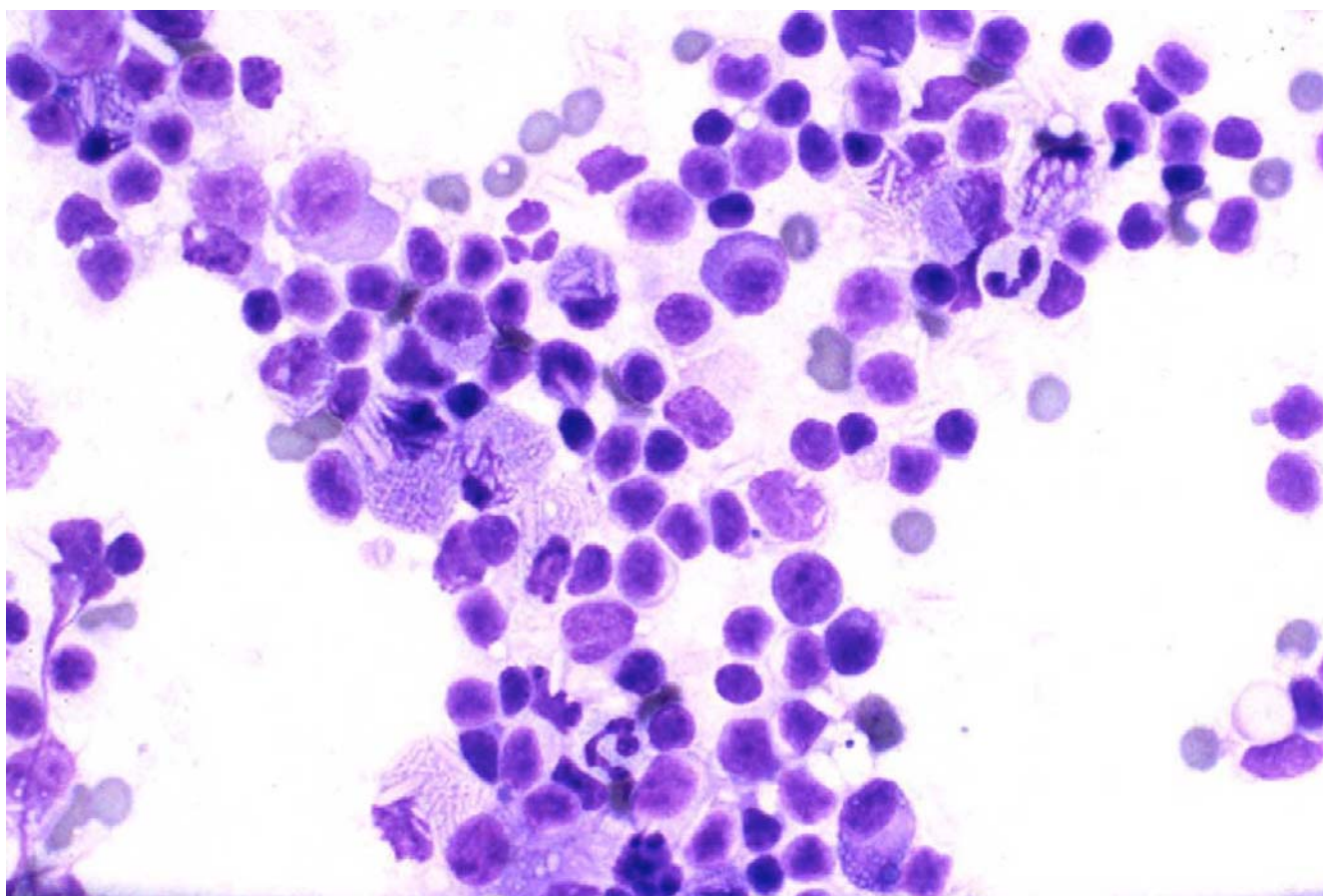


Figure 1
 Fine needle aspiration cytology of the right parotid mass showing numerous polymorphic lymphocytes. (medium high power × 550; Diff Quick stain).

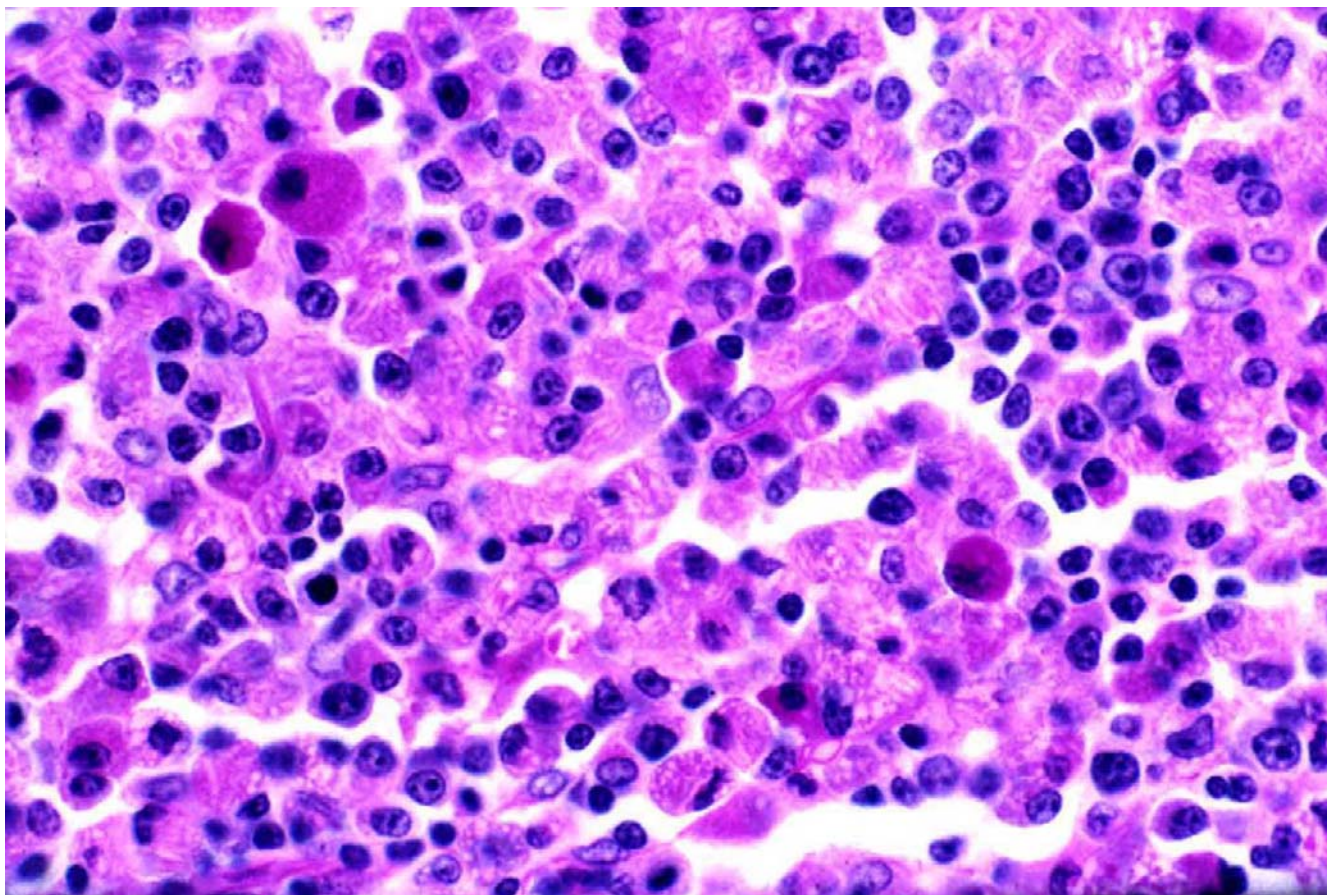


Figure 2
Crystalline inclusion bodies in tissue section with PAS stain Periodic-Acid-Schiff Staining (medium power $\times 250$) reveals sheets of neoplastic plasma cells with multiple intracytoplasmic crystalline inclusion bodies both within the plasma cells and the adjacent histiocytes.

lesional cells that were non-salivary in origin. Detailed pathological analysis showed sheets of plasma cells and plasmacytoid cells with a fair number of the plasma cells and histiocytes containing crystalline inclusion bodies many of which were linearly profiled and of multiple shapes (Figure 2, 3). Multiple cytoplasmic globules (Russell bodies), some nuclear inclusion like structures (Dutcher bodies), mild nuclear atypia and rare mitosis were also observed. Immuno-histochemical staining showed the plasma cells to be monoclonal (kappa restricted) and the small lymphoid cells in the background to be predominantly T-cells (CD45RO+), along with a smaller proportion of B-cells (CD20+) (Figure 4). A complete multiple myeloma work-up, including bone marrow biopsy, total body skeletal survey, immunoelectrophoresis, quantitative immunoglobulins and urinary Bence Jones proteins remained negative. She developed local recurrence of her disease 18 months and 30 months

later, which was managed by both radiotherapy and chemotherapy. Her clinical follow up for two years has remained uneventful.

Discussion

Solitary primary extramedullary plasmacytomas are rare and found principally in elderly people. They occur predominantly in the head and neck area with a tendency to involve the submucosal tissues of the upper airway accounting for 0.5% of all upper respiratory tract neoplasms. In two third of the cases, these neoplasms are located in the upper respiratory tract or oral cavity primarily in the nose, sinuses and nasopharynx [1].

Histopathological analysis alone is not sufficient in order to make a diagnosis of primary extramedullary plasmacytoma as multiple myeloma must be excluded by a thorough skeletal and marrow workup, immuno-electrophoresis

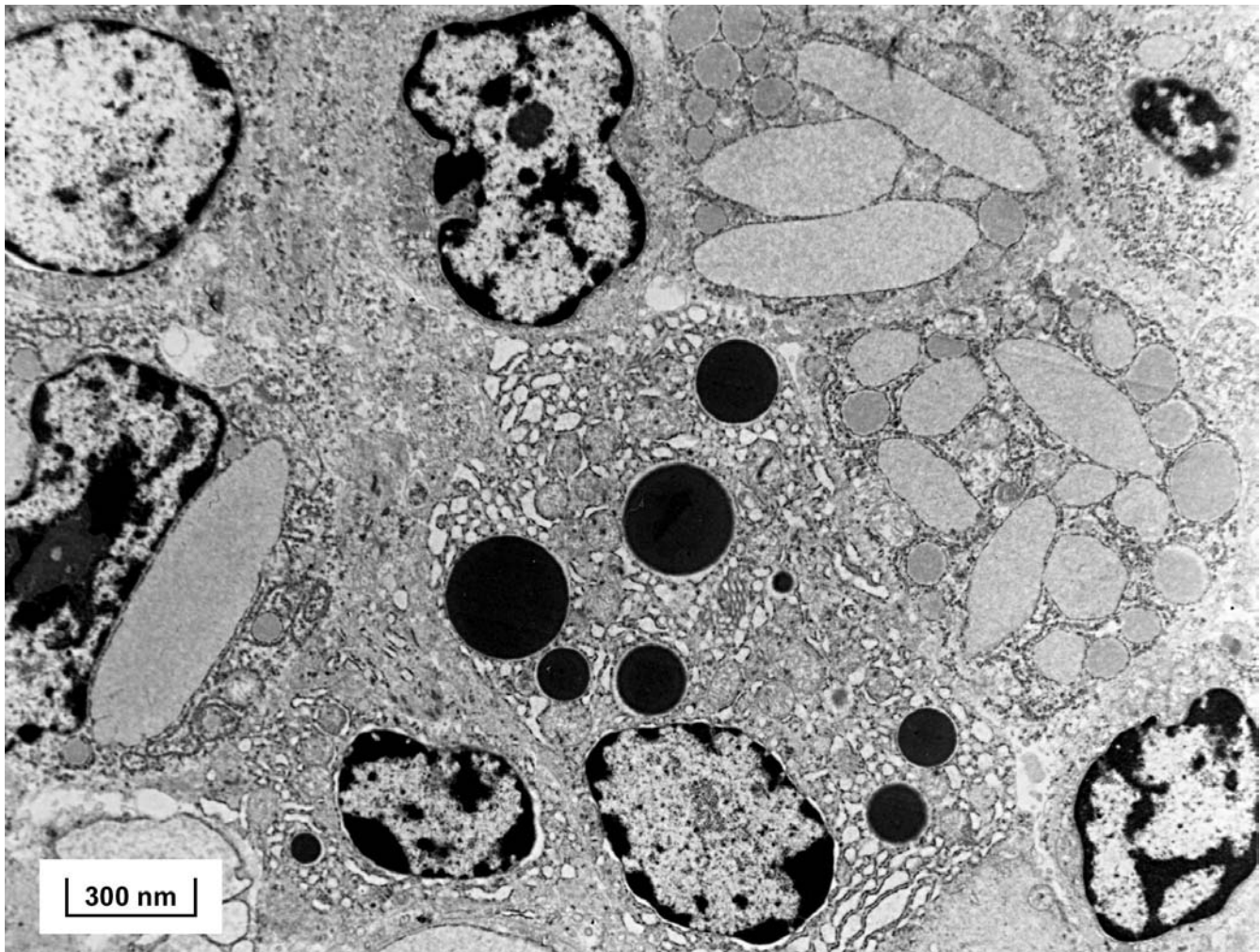


Figure 3
Electron microscopic analysis of crystalline inclusions. Ultrastructural analysis of these crystalline inclusions is demonstrated (scale 1.70 cm = 300 nm at $\times 5675$)

and quantitative immunoglobulins with absent urinary Bence Jones proteins. Fine needle aspiration cytology of plasmacytoma of the parotid gland is often reported unremarkable as in our case.

Treatment of solitary extramedullary plasmacytoma consists primarily of eradication of the local lesion and in this context surgery seems to be the primary line of treatment. Radiation therapy either alone in cases deemed unsuitable for surgery or as an adjunct with surgical removal has also been used. Local recurrences are also usually treated by radiation therapy [16] Although not proven, chemotherapy is theoretically advantageous, both in enhancing local control and in potentially eradicating sub-clinical disease to delay or prevent the development of myeloma. The pat-

terns of relapse of primary extramedullary plasmacytoma include transformation to multiple myeloma, metastasis and local recurrence. Recurrences are usually local and respond well to radiation therapy. Combined chemotherapy and radiation therapy is recommended in high-risk patients to increase the local control rate and cure rate [16] In our case radiotherapy could have been added after surgery in order to eradicate residual tumor and to prevent local relapse. However, the validity of such adjuvant therapy with complete excision of the tumor with clear margins remains contradictory. Radiotherapy was administered in our case, upon local recurrence. The second local recurrence was managed by chemotherapy.

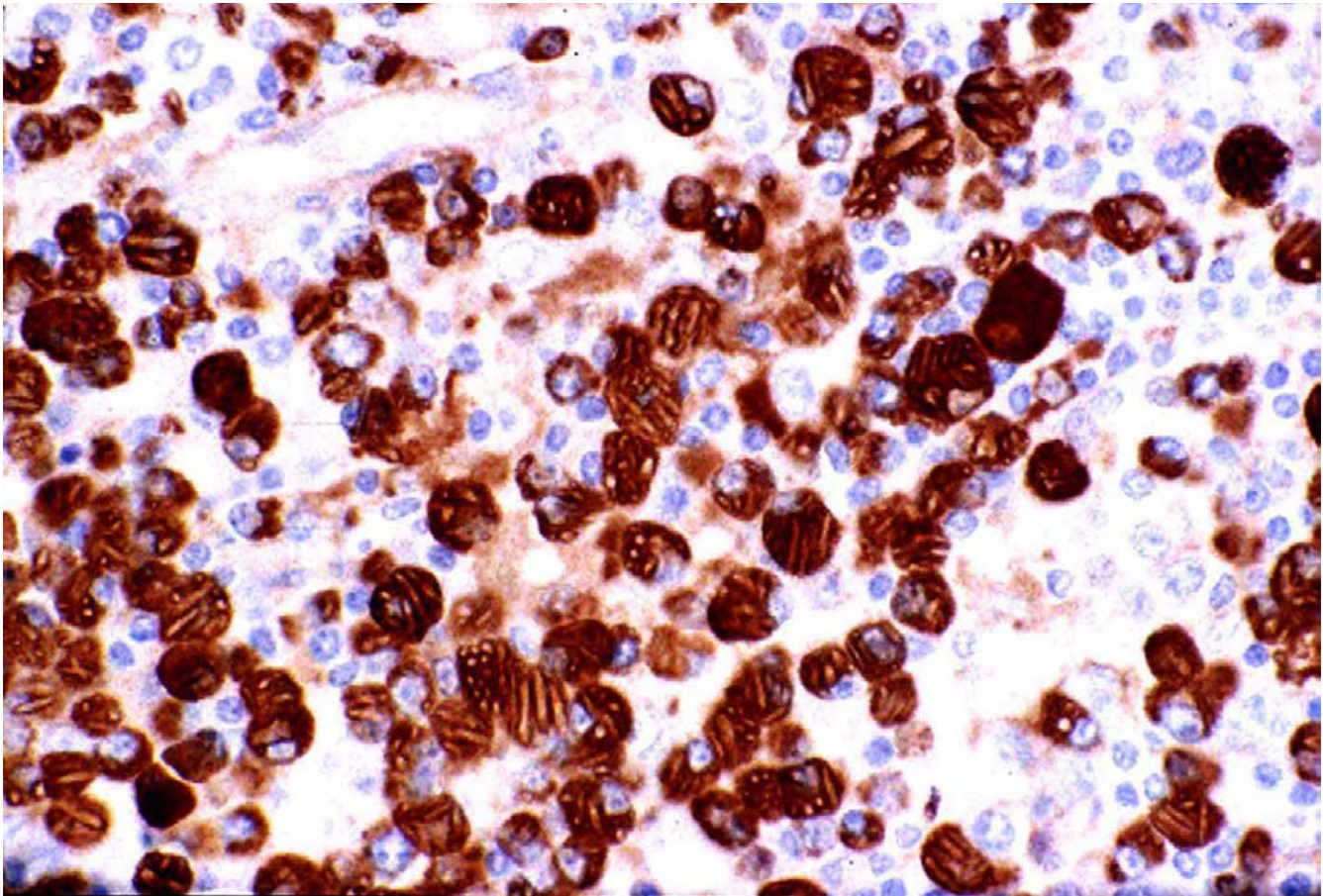


Figure 4

Immunohistochemical analysis Immunohistochemical staining shows the plasma cells to be IgG positive with kappa restriction pattern (medium power $\times 250$)

Crystalline inclusions composed of monoclonal immunoglobulins have been reported in multiple myeloma and lymphoproliferative disorders [17]. A single publication of these crystalline inclusions in primary extramedullary plasmacytoma of the parotid gland has been reported by el-Naggar et al. [11] The crystals may be rectangular or square in shape, finely uniform or globular. Crystals can be stained with anti light and/or anti heavy chains of globulin, which often manifest a light chain restriction. They can be found in histiocytes, which absorb the immunoglobulins [17]. The crystalline inclusions in our case were positive for IGg with Kappa restriction, confirming the intracellular crystallization of the immunoglobulin. Electron micrographs of the crystalline deposits show the polygonal, rectangular or diamond shape of this finding in solitary plasmacytoma. The importance of such a finding with relation to tumor progression, clinical course of the disease or prognosis in general remains uncertain.

Conclusions

Primary solitary extramedullary plasmacytoma of the parotid gland is of rare occurrence. Accurate diagnosis by fine needle aspiration requires a high index of suspicion. The presence of intracytoplasmic crystalline inclusions can further add confusion to the accurate pathological diagnosis of this entity especially at frozen section analysis.

Conflict of interest

None declared.

Author's contributions

RK is the surgical pathologist who diagnosed and followed-up this case and BT participated actively in the production of this manuscript.

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