



# IgG4 Related Cutaneous Inflammatory Pseudotumour: A Rare Case and Review of the Literature

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## Abstract

IgG4-related disease also known as inflammatory pseudotumour and include myofibroblastic tumours and plasma cell granulomas, has recently become a recognised condition. The aetiology remains unknown, with features of both allergic and autoimmune disease. We present here such a rare case and review published cases of the disease.

We present a 42 year old female patient who underwent an excision of a suspected sebaceous cyst, which was confirmed as IgG4 related disease. The patient underwent a further wide local excision due to her choice, with no evidence of recurrent disease on follow up.

IgG4 related disease can present in any organ, but occur mostly to the lung, biliary tract, pancreas and the salivary gland. The visceral inflammatory pseudo tumour manifest very differently to cutaneous manifestations of the disease, with only 17 cases reported in the English literature of this condition noted to affect the skin. The cutaneous disease behaves in a benign fashion with good outcomes from surgical excision with no current evidence in the literature of local recurrence.

This report adds another case of cutaneous IgG4 disease to the literature and reviews previously published cases.

## Introduction

IgG4 related disease or inflammatory pseudotumour (IPT) is the overarching name given to myofibroblastic tumours and plasma cell granulomas [1]. This disease commonly affects visceral organs in the thorax, central nervous system, biliary tract, pancreas, spleen, kidneys, pelvic region and salivary glands [2]. Cutaneous involvement is rare [3], with only 17 cases reported in the English literature. This report adds another case of cutaneous IgG4 disease to the literature and reviews previously published cases.

In all these cases tumour behavior has been in a benign fashion with no recurrence post surgical excision.

## Case Presentation

A 42 year old Caucasian female was referred by her general practitioner to the general surgeons with a suspected sebaceous cyst, measuring 2 cm in diameter, on her right lower flank. It was tender to palpation and noted to be discharging pus. The patient had been previously treated with several courses of oral antibiotics prior to the referral. Her past medical history included well-controlled epilepsy and asthma.

A routine surgical excision was performed under local anesthetic as a day case procedure and the specimen was sent for histology. This showed normal epidermis but within the dermis and superficial subcutaneous fat, there was spindle cell proliferation comprising blunt-ended spindle cells within pale cytoplasm, forming whorls and fascicles in a sclerotic stroma. Surrounding the spindle cell proliferation there was chronic inflammation in the form of lymphoid aggregates, some with reactive germinal centers and numerous plasma cells. No cyst was seen. The histology concluded that the features represented spindle cell proliferation with chronic inflammatory infiltrate, as an inflammatory pseudotumour such as can be seen post-arthropod bite or post a localized vasculitis (Figure 1). Immunohistochemistry revealed the plasma cells to be polyclonal. The blunt-ended spindle cells with pale cytoplasm showed some staining for CD68 and were negative for S100 and ALK-1, desmin, SMA and CD34 (Figure 2).

Following discussion at the Dermatology Multidisciplinary team meeting, the option of careful

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Received Date: 10 Oct 2016

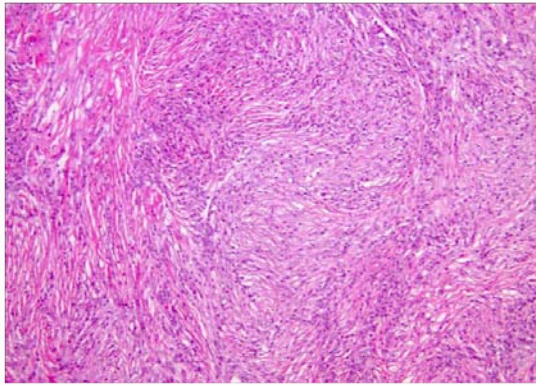
Accepted Date: 29 Nov 2016

Published Date: 07 Dec 2016

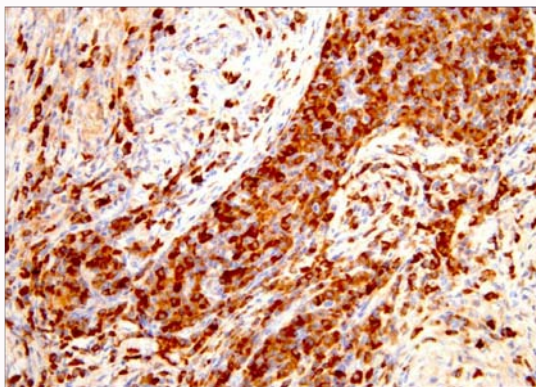
### Citation:

Kinton L, Arun A, Harmse D, Akoh JA.  
IgG4 Related Cutaneous Inflammatory  
Pseudotumour: A Rare Case and  
Review of the Literature. *Clin Surg.*  
2016; 1: 1217.

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**Figure 1:** Histology showing spindle cell proliferation with chronic inflammatory infiltrate.



**Figure 2:** Histology showing the blunt-ended spindle cells with pale cytoplasm showed some staining for CD68 and were negative for S100 and ALK-1, desmin, SMA and CD34.

observation versus a wider local excision was discussed with the patient. A wider local excision was performed successfully after four weeks from the initial surgery. Follow up from the second surgery at six weeks, showed no clinical evidence of reoccurrence and, despite the scar being sensitive, was healing well.

## Discussion

Umiker et al. [4] described the first IPT in the lung in the 1950's noting that the tumour was due to post inflammatory change rather than a neoplastic process. IPTs have since been found in many organs of the body, but pulmonary IPTs remain most common: therefore tumours are commonly classified as being pulmonary or extrapulmonary. Coffin et al. [5] in 1995 described the differences and similarities between IPT and inflammatory fibrosarcoma. IPTs have become recognized as a separate entity only in the last 20 years and despite improvements in histological and clinical examination, is still unclear whether they are due to a post inflammatory change (infection or chronic vasculitis), or a true neoplastic change. Currently the histological diagnosis of an IPT is based on the presence of polyclonal plasma cell infiltrate and fascicles of spindle cells [6].

In 2005 Arber et al. [7] found a high proportion of extrapulmonary tumours to have the Epstein-Barr virus present in them. They tested tumours originating in lymph nodes, the spleen and hepatic nodules and EBV was present in 41.2% of these tumours. Cutaneous IPT is extremely rare and only 17 cases have been noted in the English language literature [2,9-17]. The patient demographics

**Table 1:** Summary of all cases with patient demographics and anatomical sites of lesion that have been reported in literature.

Author	Number	Gender	Age	Location	Treatment
Hurt [9]	1	M	-	Arm	Excision
	2	M	-	Arm	Excision
	3	F	-	Calf	Excision
	4	F	-	Neck	Excision
Yang [10]	5	M	44	Hand	Excision
Vadmal and Pellegrini [11]	6	F	49	Arm	Excision
Nakajima [12]	7	F	25	Thigh	Excision
	8	M	89	Shoulder	Excision
	9	F	70	Neck	Excision
El- Shabrawi- Caelen [2]	10	M	15	Shoulder	Excision
	11	F	56	Arm	Excision
	12	F	56	Arm	Excision
Yung [13]	13	M	33	Shoulder	Excision
Saricaoglu [14]	14	M	57	Cheek	Excision
Pagni [15]	15	M	63	Arm	-
Gonzalez-Vela [16]	16	M	10	Arm	Excision
Su W [17]	17	M	26	Back	Excision

and anatomic locations of the lesions in these cases have been summarised (Table 1). In all but one of the reported cases of cutaneous IPT, surgical excision of the lesion has been the treatment of choice with good outcomes. For visceral disease, (more common in children) non-steroidal anti-inflammatory drug therapy has been shown to have good effect [17]. Visceral IPTs are highly vascularised and unlike cutaneous IPTs are associated with a risk of infiltration, local recurrence and rarely known to metastasise.

The paper adds another case to the growing literature on IPTs, advocating surgical resection with good margins for most cutaneous lesions. In this case the patient opted to have a wider local excision, which showed no further tumour growth and clear margins hence further resection was not required. We believe the pathogenesis and management of the condition is still evolving and not much is known about the risk of recurrence of the tumour. Though the neoplastic nature of the disease is still debated the accumulated evidence of surgical excision suggests that wide local excision is not required.

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