Methods: We carried out a retrospective analysis of the Symphony Integrated Dataverse to identify patients with advanced gout over an approximately 6-year period from October 2012 to August 2018. Patients were identified as having advanced gout if they were >18 years of age and had at least two medical claims for the diagnosis of gout on different days, separated by at least 3 months. Patients with advanced gout were identified and stratified based on their diagnosis as either chronic gout (ICD-9 274.02, 274.03, ICD-10 M1A.x) or tophaceous gout (ICD-9 274.03, 274.81, 274.82, ICD-10 M1A.xxx1). A third category designated "uncontrolled gout" was clinically defined as any patient with at least three primary diagnoses of idiopathic gout (ICD-10 M10.0x, M1A.0x) along with at least three urate tests (CPT 84550, 84560). Percent and frequency of urate testing, rheumatology specialist visits, and administration of ULT (allopurinol, febuxostat, probenecid, and lesinurad) were evaluated for each diagnostic group.

Results: We identified 191,097 advanced gout patients, including 177,610 (93%) with chronic gout, 31,475 (16.5%) with tophaceous gout, and 20,943 (11%) with uncontrolled gout. 35,992 (18.8%) were coded in more than 1 diagnostic category. The median age was 67 (range: 19-80), 24.7% were female and 75.3% male, with an average of 5.4 years of claims history in the database. As shown in Tables 1 and 2, despite the diagnosis of advanced gout, urate testing, rheumatology consults, and ULT were inconsistent. Urate testing was definitional for uncontrolled gout patients, but was done in less than 65% of patients with the diagnosis of chronic or tophaceous gout and then less than once per year. Similarly, less than 60% of subjects with advanced gout received care by a rheumatologist and less than 80% received ULT and for less than 50% of the year.

Table 1. Percentage of patients with urate testing, rheumatologist visits, and urate lowering therapy

Population	N	Urate testing (% of patients)	Rheumatologist visit (% of patients)	ULT* (% of patients)
Chronic Gout	177,610	61.72%	34.32%	76.54%
Tophaceous Gout	31,475	63.74%	57.86%	76.98%
Uncontrolled Gout	20,943	100.00%*	59.57%	81.32%

*Urate lowering therapies

*Definitional

Table 2. Frequency of urate testing, rheumatologist visits, and urate lowering therapy in advanced gout

Population	N	Mean urate tests/year	Mean rheumatologist visits/year	Mean ULT* prescriptions per patient	Mean days with active ULT* prescriptions/year
Chronic Gout	177,610	0.73	2.00	7.5	160.27
Tophaceous Gout	31,475	0.98	3.72	9	160.14
Uncontrolled Gout	20,943	2.25*	4.18	7.1	169.36

*Urate lowering therapies

*Definitional

Conclusion: The subset of patients with advanced gout, identified either as chronic, tophaceous or uncontrolled, received suboptimal care compared to current guidelines. As this group of advanced gout patients is the most affected by urate deposition, added attention should be paid to optimizing their care.

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SAT0436 CROWNED-DENS SYNDROME: A RECENT CASE SERIES IN A SINGLE CENTRE IN THE UNITED KINGDOM

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Background: The crowned dens syndrome is a rare presentation of Calcium pyrophosphate deposition disease. It is characterised by severe

acute or recurrent neck pain associated with headache, fever and elevated inflammatory markers. CT usually demonstrates calcium pyrophosphate crystal deposition in the suspensory ligaments adjacent to the atlanto-axial joint. We are reporting a series of four patients presenting with sudden onset of neck pain and raised inflammatory markers over a 9 month period.

Objectives: To raise awareness of crowned-dens syndrome, its clinical presentation, radiographic findings and management.

Methods: Four patients' electronic patient records including history, clinical findings and treatment were reviewed. Imaging was discussed at a rheumatology/radiology multi-disciplinary meeting.

Results: Four patients (2 female, 2 male) aged between 64 and 86 years presented with severe neck pain and stiffness. In all cases the onset of symptoms was less than a week prior to presentation and was associated with greatly restricted mobility. 2 patients had associated headache, arthralgia and myalgia. All patients had normal tone, power, reflexes and sensation on neurological examination. There was no pyrexia. Inflammatory markers were markedly raised with CRP ranging from 90-343 mg/L at presentation (Table 1). One patient had a neutrophilia. CT scan of the head was performed in each case and showed no evidence of subarachnoid haemorrhage, space occupying lesion or meningeal enhancement. PET-CT scan in all patients showed intense uptake around the atlanto-axial joint as well as calcification of the ligaments. All patients were commenced on a reducing regime of prednisolone starting at 40mg daily, tapering to 0mg over 6 weeks. Symptoms and inflammatory markers reduced on induction of steroids. Two patients suffered a relapse on reducing steroids and required further higher doses of steroid.

Table 1. Patient demographics

Patient	Age (Years)	Gender	CRP on presentation (mg/L)	ESR on presentation (mm/Hr)
1	64	Male	90	77
2	74	Male	247	-
3	81	Female	133	120
4	86	Female	343	112

Figure 1. Image from PET-CT scan of Patient 4 showing increased tracer uptake around the odontoid peg (arrow)

Conclusion: Patients presenting with acute neck pain, headache and raised inflammatory markers require evaluation for an infective aetiology. Pyrophosphate disease of the atlanto-axial joint however, appears to be an increasingly recognised cause of this presentation. Awareness by acute physicians, rheumatologists and radiology colleagues is important, as the condition can be misdiagnosed as Giant Cell Arteritis, Polymyalgia Rheumatica or meningitis; in order to avoid unnecessary investigations and delay in initiating the appropriate treatment. The condition is steroid responsive.

REFERENCES

- [1] Heck A, Nolan N, Rojas-Moreno C, **Crowned Dens Syndrome: Calcium Pyrophosphate Deposition Disease Masquerading as Osteomyelitis.** *J Rheumatol.* 2018 Oct;45(10):1422-1423

[2] Ledingham D, Cappelen-Smith C, Cordato D, Crowned dens syndrome. *Pract Neurol*. 2018 Feb;18(1):57-59

[3] Tagami S, Inokuchi R, Awaji K, Maehara H, Yamaguchi Y, Nakajima S, **Crowned dens syndrome and interspinous ligament inflammation due to calcium pyrophosphate deposition in an elderly man.** *Spine J*. 2016 Jul;16(7):e453-4

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SAT0437 **ABSTRACT WITHDRAWN**

SAT0438 **TO SCREEN OR “WAIT AND SEE” – CAN HLA GENE TESTING PREVENT ALLOPURINOL RELATED SEVERE CUTANEOUS ADVERSE REACTION? AND WHO ARE AT RISK?**

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Background: Allopurinol is one of the most common causes of drug-induced severe cutaneous adverse reaction (SCAR). HLA-B*58:01 gene positivity is shown to be the strongest risk factor. However, local data on HLA-B*58:01 test and allopurinol-related SCAR is scanty. Moreover, there is no consensus on routine checking of HLA-B*58:01 before starting allopurinol in Hong Kong and this test is performed according to physicians' clinical judgement.

Objectives: This study review the use of HLA-B*58:01 in daily practice, included the clinical characteristics and its implications in patients who developed allopurinol allergy in a tertiary internal medicine and rheumatology referral center.

Methods: This is a retrospective study of patients who had HLA-B*58:01 checked in Queen Elizabeth Hospital from January 2008 to December 2017. Patients' demographic data, clinical characteristics, laboratory findings, gene profile, drug allergy records were retrieved from Clinical Data Analysis and Reporting System and outcomes were reviewed.

Results: Within 10 years, 432 patients had HLA-B*58:01 checked - 23% (N=99) were positive (Figure 1). Among patients who were HLA-B*58:01 positive, 86% had clinical and/or crystal proven gout and 68% were male (M:F=67:32). Gene testing was performed as screening in 58% (57/99) and after skin reaction in 42% (42/99). Alternative urate lowering therapy was considered for patients who screened positive for HLA-B*58:01 and none developed SCAR (Febuxostat 16 patients, probenecid 3 patients). For those who reported skin reaction after allopurinol, 50% had minor rash while 50% (each 21 patients) developed SCAR. In SCAR-group, 52% was male, 76% were chronic kidney disease (CKD) $\geq$ stage 3 and/or age $\geq$ 60years, with mean age 71.2 $\pm$ 14.2 and mean estimated glomerular filtration rate 57.1 $\pm$ 30.7 mL/min/1.73m² (Figure 2 and table 1). The mean time interval to SCAR was 44.8 $\pm$ 52.1 days and the mean starting dose of allopurinol was 154.6 $\pm$ 90.7 mg/day. SCAR was associated with substantial morbidity and mortality: 71% (15/21) required steroid and/or intravenous gammaglobulin in addition to supportive care and 17% (4/21) died in the same admission due to sepsis including pneumonia. For patients who were tested negative for HLA-B*58:01, although 12% reported skin reaction, these were self-limiting and all recovered after allopurinol was stopped. Renal impairment was less pronounced in this group, yet 38% were CKD $\geq$ stage 3.

Conclusion: In our cohort, HLA-B*58:01 can identify most of the high risk patients who prompt to develop SCAR. Thus in routine practice, clinicians should consider screening HLA-B*58:01, especially in patients with CKD $\geq$ stage 3 or age $\geq$ 60year, before starting allopurinol, and consider alternatives if positive, to prevent allopurinol-related SCAR.

REFERENCES

[1] Richette P, et al. 2016 updated EULAR evidence-based recommendations for the management of gout. *Ann Rheum Dis*. 2017;76(1):29-42.

[2] Wang YH, et al. The Medication Risk of Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis in Asians: The Major Drug Causality and Comparison With the US FDA Label. *Clin Pharmacol Ther*. 2018.

[3] Khanna D, et al. 2012 American College of Rheumatology guidelines for management of gout. Part 1: systematic nonpharmacologic and pharmacologic therapeutic approaches to hyperuricemia. *Arthritis Care Res (Hoboken)*. 2012;64(10):1431-46.

[4] 4. Stamp LK, Barclay ML. How to prevent allopurinol hypersensitivity reactions? *Rheumatology (Oxford)*. 2018;57(suppl_1):i35-i41.

Figure 1. Total 432 patients had HLA-B*58:01 checked in 10 years period (January 2008 – December 2017). 99 patients were tested positive. Forty-two of these patients had the gene checked after development of allopurinol allergy. 21 of them had minor rash while 21 of them presented as SCAR. In HLA-B*58:01 negative patients, only 12% produced self-limiting rash and all recovered after allopurinol stopped.

Figure 2: Relationship between eGFR and age in patients who developed allopurinol-induced SJS / TEN / DRESS (N=21).

Table 1. Summary of patients with allopurinol-induced SCAR (N=21), 76% (16 patients) were CKD $\geq$ stage 3 and/or age $\geq$ 60 years.

Sex	Age	Indication of ULT	eGFR	Days to allergy	Allopurinol starting dose	Outcome	Treatment
F	79.6	gout	76.4	5	200mg/day	Died	IVIg, steroid
F	86.6	gout	38.5	3	unknown	recovered	Steroid
M	56.7	hyperuricemia	84.1	22	unknown	recovered	Steroid
M	55.3	gout	32.3	50	100mg/day	recovered	Steroid
M	83.6	gout	52.9	52	unknown	Died	Supportive
M	95.3	gout	13.7	237	50mg/day	Died	IVIg, steroid
F	77.4	gout	19.6	119	100mg/day	recovered	Supportive
M	72.6	gout	45.1	7	unknown	recovered	Steroid
M	73.3	gout	51.4	30	unknown	recovered	IVIg, steroid
F	74.9	gout	48.6	39	unknown	recovered	Supportive
M	75.5	gout	38.2	38	200mg/day	Died	IVIg
M	77.4	chemotherapy	91.9	18	300mg/day	recovered	Supportive
M	33.5	gout	116.6	22	unknown	recovered	Steroid
F	52.2	chemotherapy	112.5	35	unknown	recovered	Steroid
F	71.7	gout	47.3	12	unknown	recovered	IVIg, steroid
F	80.6	hyperuricemia	15.1	37	unknown	recovered	Steroid
F	68.2	gout	40.8	28	100mg/day	recovered	Steroid
M	59.3	gout	107.6	unknown	unknown	recovered	Supportive
M	81.2	gout	46	67	200mg/day	recovered	IVIg
F	81.4	gout	49	50	50mg/day	recovered	Supportive
F	57.9	gout	72.9	20	300mg/day	recovered	Steroid

Abbreviation: eGFR = estimated glomerular filtration rate (mL/min/1.73m²). ULT = urate lowering therapy. IVIg = intravenous immunoglobulin